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The American Heart Journal

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The American Heart Journal

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Original Communications

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W. F. HAMILTON, M.D., AND MAUDE E. ABBOTT, M.D.

MONTREAL, CANADA

WITH

II. A STATISTICAL STUDY AND HISTORICAL RETROSPECT OF 200 RECORDED CASES, WITH AUTOPSY, OF STENOSIS OR OBLITERATION OF THE DESCENDING ARCH IN SUBJECTS ABOVE THE AGE OF TWO YEARS.

MAUDE E. ABBOTT, M.D.

I

COARCTATION or stenosis of the arch of the aorta below the origin of the left subclavian artery has been differentiated by Bonnet in his statistical study of this subject, into two distinct groups, termed by him "infantile" and "adult" types, according to the site and form of the stenosis. His "infantile" form consists in a mere narrowing of the vessel between the origin of the left subclavian artery and the insertion of the ductus arteriosus, in the region of the fetal isthmus, and may be regarded as a persistence or exaggeration of the anatomical relations that exist before birth, when, under the conditions of the fetal circulation, the pulmonary artery carries the blood to the lower extremities through the large patent ductus, which is joined at an acute angle by the narrowed descending portion of the arch (fetal isthmus).

In the so-called adult type, on the other hand, a pathological condition exists without counterpart in normal intrauterine life,

*From the University Clinic of McGill University, and the Royal Victoria Hospital, Montreal, Canada.

Presented before the Association of American Physicians, May, 1927.

which consists of a sharp constriction, amounting in some cases to a complete obliteration of the lumen of the descending aorta, adjacent to the insertion of the ductus arteriosus either immediately at this point or just above or below it. In the pronounced cases of this type the constriction is so sudden and deep that the descending arch appears as though a ligature had been tied tightly around it at the strangulated part, and above and below this the aorta bulges outward in hour-glass fashion. In other cases there is a gradual diminution of the aortic trunk from the innominate or left subclavian, rapidly narrowing after the origin of the latter vessel to the point of constriction. Internally, the effect of the external strangulation in narrowing the lumen of the vessel is frequently accentuated by a fold or septum or diaphragm which stretches across what remains of the aortic lumen, either closing this or leaving a small central or lateral aperture, often triangular in shape, which may admit a "bristle," "probe," or "crow-quill" according to the degree and nature of the stenosis. The great vessels of the arch, especially the innominate and left subclavian arteries, with their branches, are usually much dilated as are the first three intercostals emerging below the stricture and the deep epigastric artery, for it is by means of these vessels that the collateral circulation, by which life is maintained in the extreme cases, is chiefly carried on.

Various theories have been promulgated for the explanation of this curious anomaly. That most popular in the earlier literature was first advanced by Craigie[10] (1841) in his report of a classical case in the following words. "The region of the obstruction corresponds exactly to that of the junction of the ductus arteriosus with the aorta, and the contracted point, though a little lower down, was still slightly connected with the trunk of the pulmonary artery. * * * It seems, therefore, that the obliterating action which had taken place in the ductus arteriosus had been *for some peculiar cause,* prolonged into the aorta, and had there given rise to contraction and then to obliteration of the coats of that vessel." Under the name of the skodaic theory (after the great Skoda whose support it received), this theory has been widely accepted and undoubtedly contains a germ of truth, although Brunner's[97] hypothesis, that in these cases the peculiar tissue of the ductus wall (Langer, Walkhoff) is continued into the wall of the aorta, has never been substantiated. Also, in the majority of such cases as the above, and the one described here, a mechanical factor comes into play, in that the obliterated ductus or ligamentum arteriosum persists as a thick fibrous cord, which has manifestly increased the kinking of the descending arch by exerting traction at the point of coarctation. This mechanical traction has sometimes been so strong as to produce a tent-shaped aneurysm of the aortic wall

with apex towards the ligamentum and in one case, at least (Kriegk[57]), this resulted in a double rupture of the aorta immediately above and below its point of insertion.

This explanation cannot of course apply in the few cases in which the ductus remains widely patent, and these apparently belong in a somewhat different class, in that the stenosis usually presents a certain degree of persistence of the fetal isthmus, while the upper lip of the ductus orifice may be extended inward to form a projecting shelf. Nor does the so-called skodaic theory suffice to explain the peculiar septum or diaphragm which often occupies, and may completely obliterate, the lumen at the site of insertion of the obliterated ductus, and which may be, as in the case before us, a biconcave disc, lined with clean and healthy intima of the normal aortic wall, and completely separating, at this point, the descending arch from the lower thoracic aorta. Here we are forced to accept the explanation first proposed by Reynaud[6] (1828) in his classic case, and later supported by Rokitansky (1844), to the effect that we are here dealing with a true abnormality of development arising in embryonic life, but which may, we would suggest, produce its effect at or immediately after birth. This, as Loriga[59] remarked, must have its seat in the descending limb of the primitive left aorta, which unites the left fourth (aortic) arch with the fifth and sixth arches of this side. (See Fig. 5.) The fact that this sharply localized coarctation at or near the point of insertion of the ductus has not been observed before birth, and that it is usually associated with vestigial remains of the ductus in the form of a long thick ligamentum arteriosum, would seem to indicate that the distal end of the sixth left arch (which is that part of the ductus that normally undergoes involution in utero) and the aortic wall at the entrance of this (distal end of fourth left arch) are together involved in the same process of delayed involution, and that this embryonic condition of the respective walls, together with mechanical traction by the obliterating ligamentum, may be the "peculiar cause" hypothecated by Craigie for the extension of the obliterative process beyond the ductus proper. That the fault lies at the point of junction of these three primitive arches is indicated in the present case by the anomalous vessel given off between the left subclavian and the ligamentum arteriosum (see Figs. 2 and 3), which strongly suggests a persistence of the (evanescent) fifth left arch.

ILLUSTRATIVE CASE

The patient who presented the interesting clinical features of this comparatively rare condition came into the Medical Service of the Royal Victoria Hospital in September, 1922, when he was ten years of age. He was under observation from time to time, as a ward patient, until his death at the age of fourteen. He died on the first of October, 1926, three days after his sixth admission. In the intervening four years, he had been able to attend school for the greater part of the

time. He successfully passed his grading tests for three years and at times, although against advice, he had played quite actively with his comrades. He was a very intelligent boy, usually at the head of his class. The mother stated that the boy had always been delicate; at six months he suffered an attack of measles and before reaching the age of four years he had been ill three times with pneumonia. When in his fifth year, he fell from a gallery about fifteen feet to the ground; he was carefully examined in the out-patient department of one of the city hospitals, where a few bruises and scratches gave the only evidences of injury. About six months after this fall, however, it was observed that he was not so well, and from that time on he was frequently in hospitals, on one occasion for a fractured leg due to an automobile accident. For two years as an out-patient and as a pupil in the summer school in the mountains, he enjoyed the privileges of an institute for tuberculosis where he was under suspicion as a subject of the pulmonary type of tuberculosis as well as of spinal caries. From the complaints of the patient—chest pain, a loose cough, the history of the injury, severe pain between the shoulders, weakness, etc.—it is easy to understand how tuberculosis could have been suspected. It is worth mentioning, however, that the circulatory system attracted but little attention at any time previous to 1921, when he was allowed to take up his work in the public schools.

When admitted to our service in 1922, the complaints of precordial pain, dorsal pain, dyspnea, and weakness were pronounced, while slight blueness of the surface and finger nails with clubbing of the latter were noted. Two observations practically made the chief diagnosis clear. Inspection showed at once that there was increased and very forceful pulsation in the episternal notch, in the carotids, in the subclavians, and in the interscapular areas, especially the left, as well as in the capillaries of nails and lips. The pulse in the radials was typically collapsing, while that in the femorals and tibials was absent, or at any rate impalpable. Palpation further disclosed over the pulsating area in the left interscapular region a systolic thrill, corresponding to which on auscultation one found a rough systolic murmur. Here, then, associated with other physical signs, were those indicating *aortic valvular insufficiency*, while enlargement of the small arterial branches over the upper part of the body and a total absence of pulsation in the vessels of the lower extremities pointed to a *narrowing (coarctation)* or complete stricture of the aorta, below the left subclavian artery.

A closer study of the physical signs in this case may be of interest. There was no external evidence of developmental defect. The lower extremities were in proportion to the upper and no edema was present at any time. Over the anterior aspect of the chest there was widespread pulsation more marked to the right of the sternum in the region of the second and third costal cartilages, while the apex beat was seen and felt in the sixth interspace 10 cm. from the midsternal line. A systolic thrill was widely felt in front of the chest, readily traced into the arteries over the upper part of the trunk, and felt posteriorly over those smaller arteries already described as dilated. A diastolic shock was felt over the base of the heart. Systolic and diastolic murmurs were constant, and occasionally one felt quite sure that a presystolic murmur was heard over the apex. Arterial hypertension (150/50) prevailed for the most part during the patient's visits in the hospital, with the usual high pulse pressure of aortic regurgitation, and on several occasions the pressure in the right arm was found to be higher than in the left. Fever was present on several occasions and a moderate increase in the leucocytes was observed during the early part of three of the hospital visits. Blood cultures on two occasions were negative. The blood Wassermann was negative. The x-ray pictures showed the heart much enlarged, with a bulging shadow to the right in the position usually occupied by the right auricle and superior vena cava, and complete absence

of the aortic shadow normally seen on the left of the spinal column shadow (see Fig. 1).

Among the most prominent of the patient's complaints, next to dyspnea on exertion, was that of pain over the heart and between the shoulder blades, and headaches in forehead and occiput, more marked in the erect posture. In December,

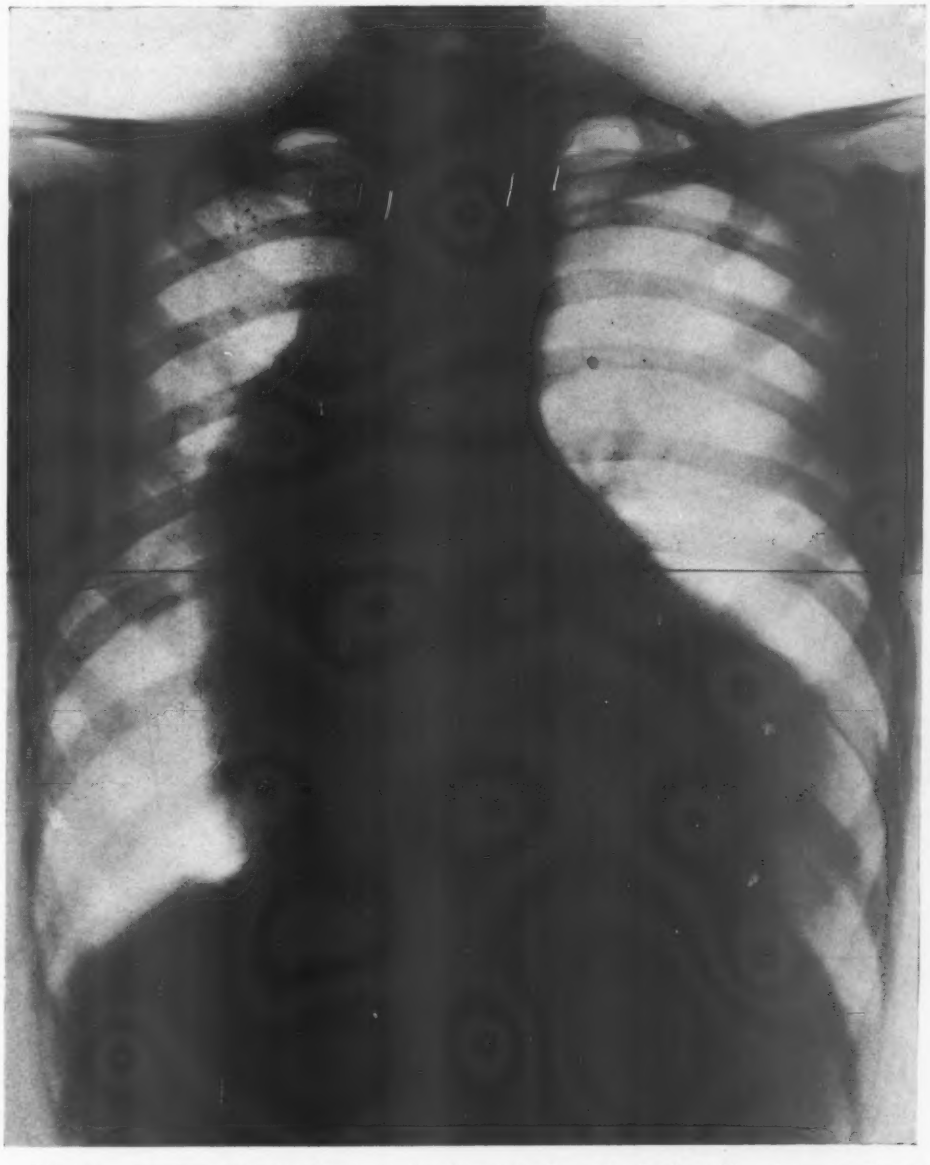

Fig. 1.—Roentgenograph taken of the heart and aorta during life of the authors' case (E. B.), showing enormous enlargement of the left ventricle, great broadening of the aortic shadow to the right marking the site of the saccular aneurysm of the ascending arch, and absence of the third left (aortic) arc indicating excessive narrowing at the point of coarctation. (From the Medical Service of the Royal Victoria Hospital, Montreal.)

1925, all these complaints were much relieved while at rest in the hospital. In February, 1926, he once more returned to school and was able to go through the term until June, with a loss of but two or three weeks.

However, in August, dyspnea supervened; he spent more time in bed. Early in September, shoulder and neck pains caused much distress, weakness and dyspnea were added, and he became feverish and bedfast. Severe pain developed over the right hip and later in the left gluteal region. The physical features described above were practically unchanged, except that the areas where the arteries were enlarged were yet more prominent. While undergoing sedative treatment for the

Fig. 2.—Coarctation of the aorta with complete obliteration of descending arch at insertion of ligamentum arteriosum, saccular aneurysm of ascending aorta with beginning dissection of wall, dilatation and displacement to the left of great vessels of the arch congenitally biscuspid aortic valve with bifid raphe, sclerosis and insufficiency of aortic valve and subaortic stenosis. Great hypertrophy and some dilatation of left ventricle and auricle. Anomalous vessels just below left subclavian artery. Large collateral circulation. Cerebral death. E. B., aged 14 years. *a*, Narrowed aortic isthmus giving off anomalous vessel. *b*, Site of complete obliteration. *c*, Ligamentum arteriosum. *d*, Interior of pulmonary artery. *e*, Enlarged periarterial gland. (From the Medical Service of the Royal Victoria Hospital, Montreal, reported by W. F. Hamilton and M. E. Abbott.)

pelvis and hip pains, the pulse became more rapid and irregular, the respirations were very difficult; the use of the right arm and leg was suddenly lost, coma developed, and death came within a few hours. The clinical diagnosis established by us fully two years before death on the basis of the characteristic symptomatology above described read: *Stenosis or atresia of the descending arch of the aorta at or near the insertion of the ductus arteriosus; aortic valvular insufficiency from*

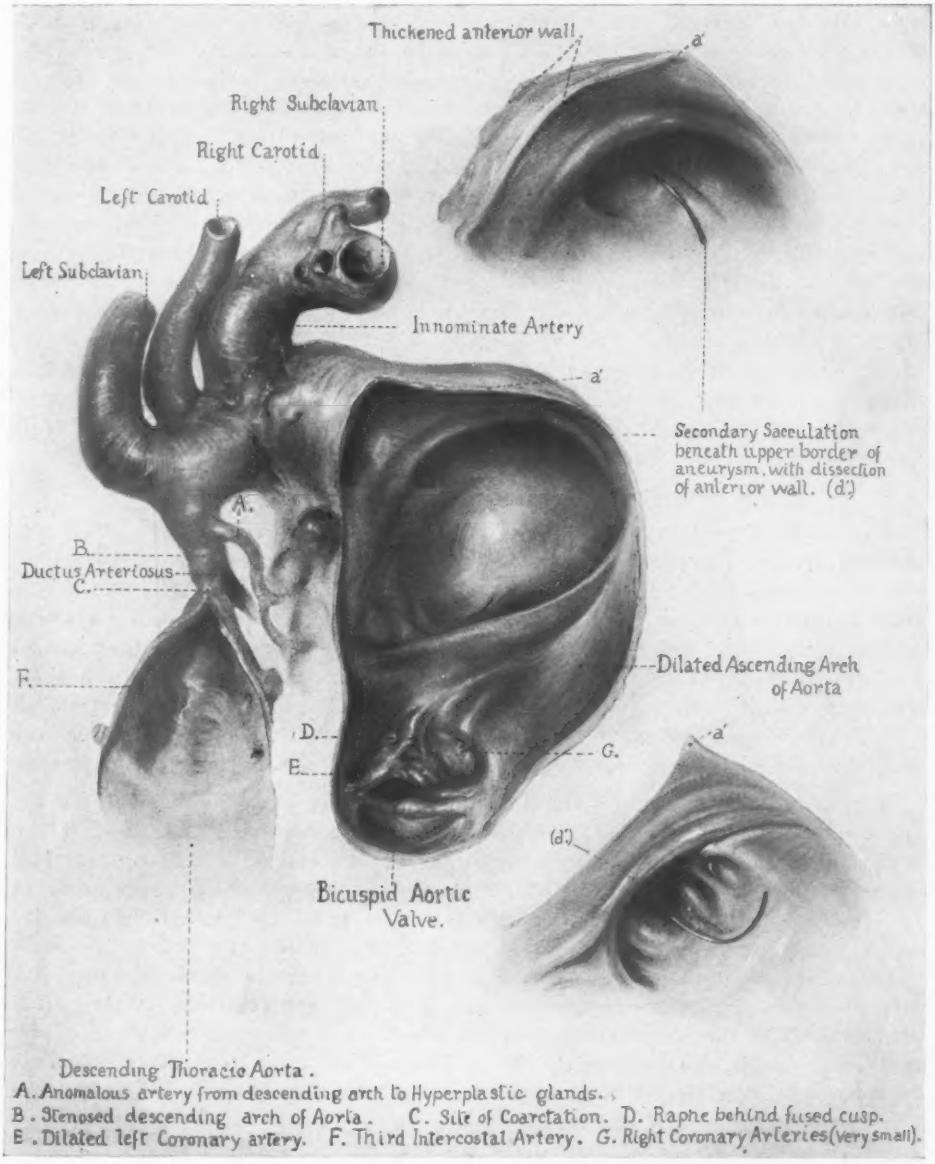

Fig. 3.—Coarctation of the aorta with stenosis of the descending arch beyond the origin of the left subclavian, and complete obliteration of its lumen at the point of insertion of the ligamentum arteriosum. Congenitally biscuspid aortic valve with bifid raphe and saccular aneurysm of right and posterior walls becoming dissecting at anterior margin. Anomalous artery from descending arch opposite left subclavian (? persistent left fifth arch). Drawing by Miss H. Blackstock (posterior view) showing the descending aorta opened from behind to reveal the obliterated area, the interior of the aneurysm, the secondary sacculation, and the bicuspid aortic valve. (From E. B. aged 14, in the Medical Service of the Royal Victoria Hospital, Montreal, published here by W. F. Hamilton and M. E. Abbott.)

chronic endocarditis, probably engrafted upon a congenitally bicuspid aortic valve; subaortic stenosis.

The autopsy showed *the aortic lumen completely obliterated at the point of coarctation*, which was exactly at the site of insertion of the ligamentum arteriosum; the wall of the vessel on either side of this presented a smooth, shining biconcave surface, suggesting that the theory of an anomalous development of the descending limb of the fourth left (aortic) arch at its junction with the sixth left (ductus), is in this case probably the true explanation of the defect; while that portion of the aorta immediately above this, as far as the origin of the left subclavian (corresponding to the fetal isthmus), is here also markedly narrowed (see Figs. 2 and 3). Other features of interest to be noted were the occurrence of a certain group of minor anomalies which are commonly associated with the adult type of coarctation, namely, a *bicuspid aortic valve*, subaortic stenosis, and an *anomalous vessel from the descending arch just above the obliterated ductus which appears to be a persistence of the evanescent fifth arch;* the evidences of a *healed infective process* leading to an extreme degree of *aortic insufficiency*, and the great hypertrophy and dilatation of the left ventricle; the existence of a saccular and *beginning dissecting aneurysm of the ascending aorta with impending rupture;* the remarkable development of the *collateral circulation;* the mode of death *apparently by cerebral hemorrhage*, which, occurring in so young a subject, suggests the existence of a congenital thinning of the cerebral arteries at their bifurcation, with the formation of a *congenital cerebral aneurysm* (Fearnsides,[248] Turnbull,[247] Parkes Weber[183]), and the presence of advanced venous congestion in all organs.

Following is a detailed description of the heart and aorta:

The pericardium was universally adherent by dense fibrous tissue. The epicardium presented punctate ecchymoses and one large hemorrhagic extravasation. The left ventricle was enormously hypertrophied and formed the apex of the heart as well as the greater part of the anterior, left lateral and posterior surfaces of the organ, while the relatively small right ventricle occupied its right lateral border. The left auricular appendix lay in its normal position, but the right appendix with the right auricle was displaced backward by the hypertrophied left chambers and the greatly dilated ascending aorta. The pulmonary artery was relatively thin-walled and of moderate size, measuring 6.25 cm. in circumference.

The aorta arose posteriorly in its normal relations, but bulged widely to the right in an aneurysmal dilatation 6.5 by 7 cm. in diameter, and then, at the beginning of the transverse arch, became reduced to a caliber of 3.25 cm., external diameter, just before the origin of the innominate artery, which vessel arose 14 cm. from the aortic cusps, having, with the other great trunks, been carried far over to the left by the dilatation of the ascending arch. Beyond it the aorta continued to diminish rapidly in caliber, becoming 2 cm. in diameter before the left carotid, and 1.5 cm. before the origin of the left subclavian artery. Immediately after the latter vessel it formed a narrow trunk 0.9 cm. in diameter and continued to decrease gently for a distance of 2 cm. when it received on its inner concave surface a thick cord 0.4 cm. in diameter, which is the ligamentum arteriosum or obliterated ductus. At this point the descending arch presented on its outer surface a shallow ring of further constriction, which coincided internally with a complete obliteration of its lumen, the aorta above and below this point ending blindly in a shallow biconcave disc lined on both sides by shining intimal tissue. About 1 cm. above the insertion of the obliterated ductus, and just opposite the origin of the left subclavian an anomalous artery about the size of a large knitting needle arose and passed down to a mass of enlarged glands on the right side of the arch and was there lost (persistent left fifth arch?). Immediately below the seat of obliteration the aorta widened to an internal circumference of

4 cm. The descending thoracic aorta just below the atresia showed several small patches of atheroma. The lumina of the first three intercostal arteries appeared dilated. The descending aorta narrowed rapidly, measuring 2.3 cm. in diameter at the lower border of the eighth rib, and 2 cm. after the origin of the celiac axis. The collateral circulation had been carried on by the tortuous and hugely dilated internal mammary arteries (see Fig. 4) with the deep epigastrics, and the superior or first intercostals, dorsalis scapulae, and other branches of the subclavians, with the upper aortic intercostals.

On laying open the ascending aorta its interior wall was found to present a marked hyaline thickening of its outer coats (0.6 cm.) and to a lesser degree of the intima (0.2 cm.) making a total thickness of 0.8 cm., and it was occupied

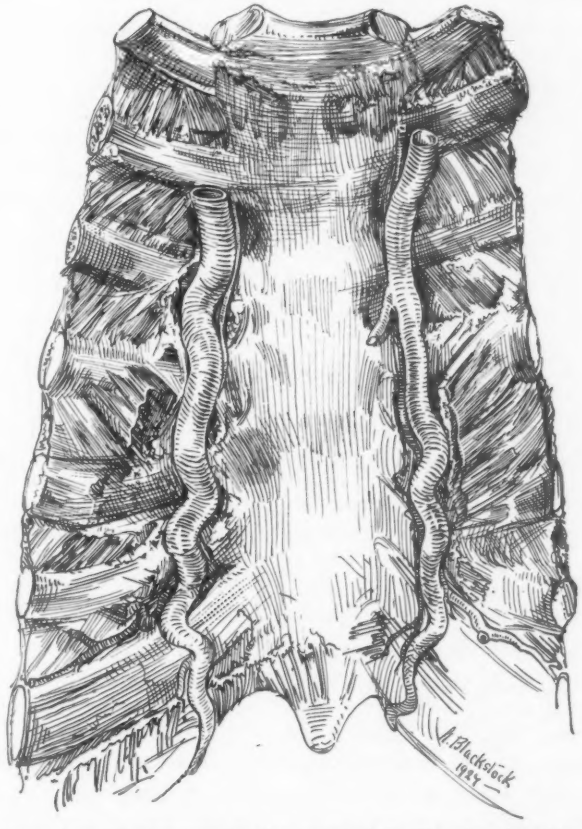

Fig. 4.—Interior view of sternum in the case of E. B., showing the great dilatation and tortuosity of the internal mammary arteries as a result of the enormous development of the collateral circulation through these vessels. (Compare the similar picture in Dr. Parkes Weber's case.) (Drawing by Miss H. Blackstock from the case in the Medical Service of the Royal Victoria Hospital, Montreal, published here by W. F. Hamilton and M. E. Abbott.)

on its right side by a large saccular aneurysm with an orifice 5 cm. wide, with thin shelf-like margins, which concealed a thick fenestrated base that was tunnelled out beneath the upper border into a deep secondary sacculation with beginning dissection of the anterior wall. The posterior wall was smooth and elastic, with patches of atheroma and early calcification.

On laying open the heart the interior of the left ventricle was found to be lined by a greatly thickened endocardium, especially over the septum and posterior wall about the insertion of the anterior papillary muscles; the aortic segment of the mitral valve presented on its ventricular surface a large patch of sclerosis,

and there was a sclerotic ring (subaortic stenosis) about 0.5 cm. below the right aortic cartilages and cusps. The aortic segments were much thickened along their free borders and presented hard gristly excrescences, aortic insufficiency existing. The right posterior aortic cusp was elongated, and pouched, and measured 3 cm. along its free border, while the anterior and left posterior (coronary) cusps formed a single segment 3.2 cm. long, behind which lay a low broad raphe, which incompletely divided the floor of the sinus into two nearly equal parts, and was inserted by two thick chordae into the aortic surface of the combined cusp along its free border, the one into the back of its thickened and fused corpora arantii, and the other 0.5 cm. to the left of this. The minute orifices of a very small right coronary artery and of an accessory coronary artery lay behind the right half of this united (or incompletely divided) segment, and a very large left coronary orifice occupied the extreme posterior angle behind its left half. The aortic ring measured 8 cm. in circumference but the valvular orifice only 6 cm. and was further narrowed by the rigidity of the anomalous cusp.

The cavity of the left ventricle was enlarged, measuring internally 11 cm. from the insertion of the aortic cusps to the apex, but its increase had not kept pace with that of the wall which was enormously hypertrophied, measuring 3.5 cm. at its thickest part. The left auricle was also markedly hypertrophied (0.6 cm.) The foramen ovale was closed. The mitral orifice was 6.5 cm. in circumference, and its valves were thickened. The right auricle was dilated and the caval orifices were very wide. The tricuspid valve was normal and its orifice 11 cm. in circumference. The right ventricle was very small, but its walls were moderately hypertrophied (0.1 cm.). The wall of the superior cava and roof of the right auricle were firmly adherent to the postero-inferior surface of the aneurysmal sac.

Summary of Anatomical Diagnosis of the Heart and Aorta

Coarctation of the aorta with stenosis of the isthmus and complete obliteration of lumen at point of insertion of ligamentum arteriosum. Diffuse dilatation of ascending arch with saccular aneurysm of right anterior wall and secondary sacculation along upper border with beginning dissecting aneurysm. Marked dilatation with tortuosity of subclavian, internal mammary and dorsalis scapulae arteries above, and of upper three intercostals below the seat of coarctation, and of deep epigastric arteries, with establishment of extensive collateral circulation. Anomalous artery from descending arch just below left subclavian and above obliterated ductus. Bicuspid aortic valve with partly obliterated and fenestrated raphe and shortening of incompletely divided cusp. Chronic sclerosing endocarditis of aortic valve with aortic stenosis and insufficiency, and sclerosis of mural endocardium of left ventricle, with subaortic annular thickening below aortic cusps. Great hypertrophy with some dilatation of left ventricle and left auricle. Slight hypertrophy of right ventricle, dilatation of right auricle. Completely adherent pericardium, epicardial ecchymoses, and subepicardial hemorrhagic extravasation.

Microscopic Examination (Report by Prof. T. R. Waugh).—Sections were taken from the ventricular wall, ascending aorta, aneurysmal sac, and descending aorta, and stained by hematoxylin and eosin, Mallory's connective tissue, and Weigert's elastic tissue method.

Sections of the *myocardium* showed moderate hypertrophy of some of the muscle cells; moreover, the fibers appeared widely separated and the connective-tissue supporting substance was accentuated. There was no evidence in the sections examined, however, of any productive inflammatory changes.

Sections of the *ascending aorta* showed a considerably thickened wall which was the seat of a more or less diffuse hyaline fusion of the cellular elements. This was less marked in the intima, but very conspicuous in the media which was fused

into a practically homogeneous, pale-staining substance. The elastic fibers of the intima appeared stretched, broken and irregular, while those of the media were entirely lost. The adventitia was relatively free and showed no evidence of inflammatory reaction.

The same general description holds for the *aneurysmal sac*, except that the wall was here much thicker, the outline and separation of the various coats was indistinct, and the elastic tissue of the intima was not as well preserved.

Sections of the *descending aorta* showed the wall to be very thin, hypoplastic, and atrophic. Differentiation of the coats was indistinct. The elastic tissue stains showed a large number of elastic tissue fibers throughout the intimal and medial coats.

REMARKS

Coarctation of the aorta of the type here described is essentially a condition that threatens persons in the prime of life, between the ages of twenty and forty years, which are the years of greatest economic efficiency. That is to say, in the great majority of the cases, the condition remains latent so far as symptoms are concerned, the gradual development of the collateral circulation keeping pace with the demands of the organism, and the potential strain that may thereby be thrown upon the heart and arterial walls remaining in abeyance, until some intercurrent infection has weakened the myocardium, or some excessive muscular exertion has increased the hypertension in the upper part of the body beyond the limits of the circulatory reserve. From the point of view of symptoms, therefore, we may distinguish three sets of cases:

a. Those in whom the condition is apparently entirely latent throughout life, as in Reynaud's remarkable patient who reached the age of ninety-two years with the development of an enormous collateral circulation but no hypertrophy of the heart. In such cases the condition will usually reveal itself at the autopsy as a "*surprise d'amphithèatre*," either an accidental finding, or as the cause of sudden death from rupture of the heart or aorta or other artery, in an individual who has been apparently in perfect health until a few hours or days before his decease.

b. In a previously robust and vigorous person in the full tide of early adult life, signs of cardiac insufficiency may suddenly set in as the result of what would otherwise be a quite inadequate cause, and pass rapidly or gradually into a state of complete decompensation, followed by cardiac death. In the words of Professor Meixner, "It is the unavoidable wear and tear of life, which has little effect upon the majority of other people, that comes into action here. The subjects of this malformation are seriously affected, and probably it is only the few who are protected from such injuries as the common diseases of childhood and of other periods of life, who become old."

c. In a few cases, as the one here reported, signs of vascular surcharge in the upper part of the body, with dyspnea and palpitation as evidences of cardiac fatigue, may exist from earliest childhood, rendering the diagnosis and the precarious condition of the individual plain.

Special Features of This Case.—The patient whose condition is here communicated, falls into the third clinical category above enumerated, in that signs and symptoms of the aortic obstruction were present from an early age. The grave changes that had occurred in the myocardium and the gross deformities of the aortic valve, with the early history of an infective process, supply ample ground for this early onset of the clinical syndrome, which has been fully discussed above. The abrupt termination at the age of fourteen was, however, the result of another set of causes which appear to be in a sense interdependent, and which together supply a classic picture of the clinicopathological aspects of the anomaly itself, which is nowhere surpassed in the literature. We refer to the bicuspid aortic valve complicating a complete atresia in the course of an otherwise perfectly healthy descending arch, the strong ligamentum arteriosum, which, by its attachment to the point of atresia and to the pulmonary artery may, as Thoma pointed out, exert some counterstrain on the ascending aorta and so be a factor in the rupture that so often occurs; the aneurysmal dilatation, dissection, and impending rupture of this trunk that had taken place in this case in this situation; the extensive collateral circulation in the upper part of the body, which must have supplied, in its terminal ramifications in the brain and elsewhere, a *locus minoris resistentiae* under the high intraarterial pressure that existed here, and the cerebral death. In addition, the vascular phenomena of dilated and tortuous, visibly pulsating, peripheral vessels perceptible over the thorax, with lessened pressure and pulsation in the arteries of the lower extremities; the signs of left-sided hypertrophy and the typical x-ray picture of absence of the left fourth (aortic) arch, with broadened shadow on the right side made by a greatly dilated ascending aorta (see Fig. 1), presented the cardinal diagnostic signs of this condition in exquisite pronouncement. Further comment upon these and other features of this remarkable case may best be made in discussion of the other similar ones in the literature which are reviewed statistically below.

II

Statistical Study and Historical Retrospect of 200 Recorded Cases, with Autopsy, of Stenosis or Obliteration of the Descending Arch.

Maude E. Abbott, M.D.

A careful revision of the cases abstracted in the earlier surveys made of this subject by Craigie,[10] Eppinger,[48] Kriegk,[56] Barie,[78] and Bonnet[101] (1903) has yielded 101 cases, confirmed by autopsy, of

stenosis of the descending arch of the aorta without grave complicating anomaly in subjects over three years of age (i.e., in "adults"). I have abstracted 35 more such cases for my monograph[204] in Osler's System in its three successive editions of 1908, 1915, and 1927, making a total of 136* cases collected from the literature to the latter date. With the help of the bibliographies recently compiled by Drs. J. T. King[208] and L. M. Blackford,[209] Prof. K. Meixner[175] and other sources, I have collected 47 additional cases from the old and recent literature, making a total of 183 such cases, which have been available to us, mostly from the original sources, for personal study (see Chart II). In addition, through the kind cooperation of Dr. L. Minor Blackford,[209] of the Mayo Clinic, data from 17 other "new" cases, collected by him from the literature from sources not at present accessible to me, have been supplied to me for the purpose of these statistics from

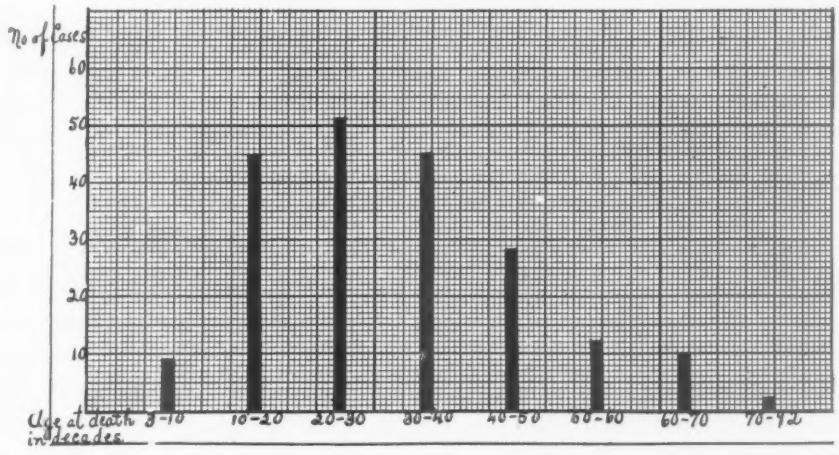

Chart I.—Average age at death of 200 cases of coarctation of the aorta in adults with autopsy from the literature.

his valuable article on this subject (now in press). This brings the total number of cases collected at the present time and here reviewed to 200, of which 64 are "new" cases, in that they have not been included in any of the published series above mentioned. Of these, 17 as previously stated, are in Blackford's series only, 41 are common to both this and his article, and 6 (cases by Oehl,[139] Taruffi,[140] Wadstein's Obs., 96,[144] Hinrichsmeyer,[166] Nieuwejaar,[178] and Libman[180]) appear in this series only. In addition we know of five other uncomplicated cases with autopsy in adults as yet unpublished. One of these is in Dr. Blackford's Mayo Clinic series, and is reviewed by him

*The number of cases of coarctation with autopsy in "adults" collected to that date was mentioned in my article in Osler's System, 1927, iv, 774 as being 155. The additional 18 cases there referred to are of a relatively slight grade of coarctation complicating other more marked anomalies, which therefore dominated the clinical picture and were classed as the "primary" lesion. These have been excluded for the purpose of this communication, but are listed below in the discussion upon the incidence of associated anomalies.

in his thesis; one is a specimen in the McGill Museum from an elderly man with a moderate degree of stenosis at the ligamentum arteriosum, and aneurysmal dilatation with great atheroma of the ascending aorta; and two are from the autopsy records of the Massachusetts General Hospital in men aged, respectively, forty-nine and twenty-nine years; data from whose cases have been kindly supplied to us by Dr. Paul D. White. "In a very careful search recently made by Dr. George Houck into the records of 5000 consecutive autopsies at this hospital, there were found only these two cases of definite coarctation of the aorta, and two other doubtful cases. Apparently none of the cases showed any symptoms of limitation of cardiac function from the coarctation. One of them had visible pulsation of the arteries." (Personal communication from Dr. White.)

The fifth case was presented before the American Association of Anatomists in May, 1928, and will be published in their Transactions by Dr. A. G. Carmel, of the University of Cincinnati. In a man aged 40 who died of acute cardiac dilatation following acute alcoholism, superficial tortuous arteries were observed at dissection over the trapezii muscles, and the aorta was found to be constricted to a circumference of 2.2 cm. Immediately below this it widened to 6.5 cm. and gave off here a number of tortuous branches. An interesting collateral circulation had developed.

ANATOMICAL FEATURES

As is evident from the descriptive list given below, the great majority of these cases in subjects over two years are more or less typical examples of the so-called "adult type," in which, sometimes suddenly, but usually after some preliminary narrowing, a sharply localized abrupt constriction occurs, situated *at* (in 90 of the 183 cases studied by us), or *immediately below* (in 43 of the other cases), the insertion of the ductus arteriosus or ligamentum arteriosum, and which is produced internally by a septum or diaphragm formed of the inner coats of the aorta occluding, or partly occluding, its lumen, and externally by a sharp inward kinking and annular thickening of the wall having the effect of a ligature. In 19 cases of this series the exact seat of the stenosis was not mentioned, and in 18 others it was spoken of indefinitely as "below the left subclavian" without specifying its relation to the ductus. In 6 cases, however, those by Wrany,[52] Scheiber,[44] Babington,[102] Pappenheimer (Case 2[118]), Nixon,[8] and Knierim,[141] it was definitely stated to lie "*above the ductus*" and in 7 others it was described as lying *at* (cases by Pelletier,[11] Fawcett [Case 12[112]], Woltman and Shelden) or *above* the origin of the left subclavian (Fraenckel's Case 2[156] and those by Oehl,[139] Maigne,[60] and Lesseliers[142]). In both the latter case and that of Fraenckel the stenosis lay immediately beyond the left carotid, and the subclavian arose distal to the stricture, although its orifice was involved in it. In these cases, which are rightly classed by Blackford as "*atypical*," the ste-

nosis lies quite out of the proximity of the ductus, and the involution of this cannot therefore have had any direct bearing upon their causation, a point which renders them of much interest from the standpoint of etiology. In Lesseliers' case, a farm girl aged twenty years, with no history of rheumatism or other serious illness, who died with cardiac decompensation of rapid onset, the arch was constricted as though by a ligature to a caliber barely admitting the tip of the little finger just beyond the left carotid, thereby narrowing also the mouth of the left subclavian. The ascending aorta up to the point of stenosis was the seat of extensive "atheroma," and the aortic valve was also diseased, but the anomalous condition of the latter (insertion of an abnormally small cusp) and the ligature-like constriction of the arch externally suggest a congenital origin of both deformities.

State of Ductus Botalli.—In 7 cases of the 183, the coarctation took the form of a diffuse narrowing of the "isthmus," between the left subclavian artery and the ductus; that is, it was a persistence of the infantile type into adult life. In 4 of these cases, namely, those of Chevers,[21] Peacock,[103] Almagro,[65] and Fawcett's Obs. 11,[111] the ductus was widely patent; in two others, by Bronson and Sutherland[163] and Laubry,[179] it was obliterated; and in one, that by Kohn,[152] this point was not mentioned, as in 66 other cases of the 183 examined.

In 11 other cases (making 15 in all among the 183 cases) the ductus was noted to be patent in its entire length. These were those reported by Nixon,[8] Hamernjk (Case 3[40]) (admitting a boar-bristle), Redenbacher,[53] Viaud-Grandmarais,[64] Hornung,[74] Babington,[102] Peacock,[103] Fawcett[111] (Obs. 11, and Obs. 15), Horder,[151] Stauing,[157] and Meixner.[175] In addition the ductus was patent in 3 of Blackford's 17 cases, in those by Hansteen,[199] Lantinga,[185] and Kovesi,[190] making in all 18 cases of patency among the 200 of coarctation in adults, which is an incidence of 10 per cent.

In the remaining 101 cases of the 183 accessible to us the ductus was not patent. In 79 of these it was obliterated and usually ligamentous, forming a thickened cord 1 to 1.5 cm. or even (in one of Eppinger's cases) 2.9 cm. long. This solid cord was in some instances lost in the thickened wall of the aorta at the point of constriction, and in others its attachment corresponded with a funnel- or almond-shaped depression (as in Laennec's early case), forming a true "traction aneurysm" of the wall which was sometimes of large size, and occasionally ruptured, as in the cases of Kriegk[56] and Willigk.[41] In 21 other cases the ductus was stated to be "closed," but in 5 of these it was open on the pulmonary side (as in Fletcher's[16] case in which it admitted a bristle but was closed on the aortic side by a thin membrane), and in 8 others it was open on the side of the aorta (as in Barker's[39] case where it

formed a diverticulum the size of a goosequill, closed on the side of the pulmonary artery). In the remaining case of the 101, that by Hale White,[77] no trace of the ductus was found on careful examination.

The degree of stenosis varies from a complete obliteration or an "extreme" narrowing to a relatively slight reduction of the lumen of the aorta at the point of stenosis. Among the total 200 cases there were 47 of complete atresia, 108 of "extreme" and 45 of "moderate" stenosis. In order to present a convenient oversight of the whole, these cases are listed below in these three groups in the order of their date of publication.

Following are the 47 cases of *complete atresia:*

Graham[2] (1814), complete obliteration as though by a ligature below (closed) d.a.**; Jordan[7] (1830), aorta completely obliterated 3 lines below obliterated d.a., and ending in a culdesac above and below the occlusion; Craigie[10] (1841), sudden contraction of descending arch with complete obstruction just below obliterated d.a.; Roemer[14] (1839), aortic caliber entirely obstructed for half an inch below d.a.; Wise[17] (1842), below left subclavian, aorta sharply constricted as though by a ligature, becoming completely closed at level of obliterated ductus; Hamernjk[20] (1844), marked circular contraction forming transverse septum just at d.a., completely closing aortic lumen; Hamernjk-Oppolzer, Case 2[25] (1848); West*[137] (1848), aorta appearing as though ligatured by a well-defined constriction at and immediately below the ligamentum arteriosum, completely obliterating its lumen; Lebert[27] (1852), double stenosis by transverse septum *above* d.a. leaving opening 0.25 mm. in diameter and below this, *at* ductus, a second septum obliterating lumen; Rokitansky-Loebel[31] (1853), marked stenosis 1½ lines beyond origin of left subclavian with complete obliteration above and below ductus; Rokitansky-Dlauhy[29] (1856), stenosis at insertion of ligamentum arteriosum to 1 line in diameter, with complete occlusion here by atheromatous plaque; Harrison*[138] (1855), aorta just below the arch forms an obliterated cord 3 inches long; Jones[34] (1856), aorta forms a ligamentous cord ½ inch long; Wood[37] (1858), constriction like a piece of packthread tied tightly around vessel; Scheiber[44] (1863), aorta obliterated 2 lines *above* ductus; Liouville[81] (1869), aorta obliterated a little below ductus; Bradley[82] (1871), sudden constriction as in tying a sausage, completely obliterating lumen, with normal caliber immediately below; Hornung[74] (1876); Luttich[72] (1876), aortic arch ends in a culdesac which is united with the descending aorta by a ligamentous cord at level of obliterated d.a.; Legg[85] (1878); Sommerbrodt[75] (1883), aorta forms dense fibrous cord at obliterated d.a.; Manneberg[86] (1884), aorta ended blind below obliterated d.a. in a solid segment ½ inch long, shaped like a collar; Paltauf[87] (1887), funnel-shaped atresia 0.5 cm. long, behind d.a.; Martens[90] (1889), double stenosis at upper and lower ends of dilated isthmus, with obliteration of lower one at ductus by folding in of walls to form a thin occluding membrane, marked externally by a deep angular groove on convex side of arch; Decker[91] (1890), hour-glass shaped obliteration just below closed ductus; Brunner[97] (1898), obliteration as by a septum internally and ligature externally at obliterated ductus; Dickinson and Fenton[98] (1900), isthmus narrowed by a sharp ridge, below this complete obliteration; Haberer*[148] (1903), isthmus 13 mm. wide and 15 mm. long, terminates blindly in a convex surface which appears directly continuous with the ligamentum arteriosum, descending aorta attached almost at a right angle;

**d.a. = ductus arteriosus.

*New series in this and Blackford's article.

Fawcett, Obs.†3[108] (1905), funnel-shaped stenosis after left subclavian with complete occlusion just below patent d.a.; Strassner†[122] (1909), narrowing after great vessels with complete obliteration at d.a.; Sella, Case 2†[124] (1910), the atresic aorta formed a solid cord 1.2 cm. long, uniting blindly with the descending aorta at attachment of ligamentum arteriosum; Moon†[126] (1912), narrowing after left subclavian with complete occlusion after closed d.a.; Fraenckel,*[155] Case 1 (1912), aortic arch narrows and ends blind, just below ligamentum arteriosum by a deep constriction; Kolisko, Case 2*[160] (1913), aorta forms hard obliterated cord ½ cm. long just behind ligamentum arteriosum, which appears like its continuation; Kolisko, Case 3‡[161], complete closure at isthmus; Lecount†[127] (1913); Oppenheim*[164] (1918); Hart, Case 1*[167] (1920), hour-glass constriction completely occluding lumen at obliterated ductus; Loeper et Marchal†[130] (1922), constriction as by a ligature below d.a.; Meixner*[175] (1922), funnel-shaped with complete closure 2 cm. below left subclavian with immediate dilatation beyond; Strassman, Case 5*[171] (1922); Strassman, Case 8*[174]; Bahn*[177] (1925), externally no constriction but on laying it open there is directly under the insertion of the ligamentum arteriosum a complete diaphragm occluding it, and a narrowing above and below this; Hamilton and Abbott†[136] (1926) (this case) after narrowed isthmus the aorta ends blind in biconcave disc, at insertion of ligamentum arteriosum; Woltman and Shelden*[182] (1927), aorta narrowed rapidly and ended blindly at origin of left subclavian (*above* obliterated ductus), and Koster-Forselius§[196] (1915), and Zenoni§[193] (1911), from Blackford's own series.

In 108 other cases of the 200, the stenosis was "extreme," that is, the aorta at the point of constriction, although permeable, was reduced to a very small lumen, not larger than a crowquill or pencil (6 mm.) and frequently so minute as to allow only the passage of "a hair" or "a bristle." Following is a descriptive list of these cases of *extreme stenosis:*

Paris[1] (1791), a stenosis "the diameter of a writing pen"; Otto[4] (1824), scarcely admitting a pen; Meckel[5] (1827) "admitting a fine straw"; Reynaud[6] (1928), admitting a quill; Legrand[9] (1835), an opening in a circular septum 1½ lines in diameter; Pelletier;[11] Cruveilhier,[12] admitting a fine stylet; Mercier;[13] Hargrave,[15] a sudden stenosis ½ inch long admitting a goosequill; Andral[61] (1840); Tiedemann[19] (1843), a cartilaginous septum ½ inch long presented at the attachment of the obliterated ductus an opening ½ line in diameter; Bochdalek[22] (1845); Dubreuil, Case 2[80] (1847); Muriel[18] (1847); Crisp, Case 2[63] (1846); Hamernjk-Oppolzer[25] (1848), Case 2; Hamernjk-Oppolzer, Case 3;[40] Van Leuven[26] (1849); Barth[28] (1852), a thin diaphragm formed of thickened aortic intima was pierced by an opening 2 lines in diameter; Rokitansky, Obs. 20[30] (1853); Dumontpallier[33] (1856), the aortic isthmus was reduced to 13 mm. in diameter and its lumen was almost occluded at the ductus by a diaphragm formed of aortic media and intima, the outer coat passing over it like a bridge, and pierced by a minute triangular opening 0.012 mm. in diameter; Rosenstein[43] (1857); Leudet[35] (1858); Wilks[36] (1859) constriction at d.a. as though by a ligature admitting fine probe; Viaud-Grandmarais[64] (1857), aorta ligatured as though by a thread below left subclavian; Kjellberg[38] (1860); Barker[39] (1860); Almagro[65] (1862); O'Flaherty[50] (1868), double stenosis, the lower at d.a. admitting a crowquill; Pommier[67] (1868); Degen[47] (1868); Eppinger[48] (1871) Case 1, stenosis produced by a projecting ridge barely admitting a sound; (Case 2[49]); Traube[51] (1871); Wrany[52] (1871), callous thickening projects into aorta permitting

†Series in Abbott's monograph, Osler's System.

passage of a goosequill; Riegel[68] (1872); Forster (1854), Case 1;[45] Case 2;[46] Redenbacher[53] (1873); Erman[54] (1873); Stoll-Krotowski[69] (1873); Moutard-Martin[70] (1874); Goodhart, Case 1[71] (1874), Case 2;[83] Taruffi‡[140] (1875); Moutard-Martin[73] (1876); Kriegk[56] (1878), Case 1, a resistant ring forming a cuff around the aorta 0.5 cm. long, and admitting only a bristle; Case 2,[57] annular constriction producing a ridge internally leaving lumen 0.038 cm.; Knierim*[141] (1880), stenosis as though by a ligature 2.5 cm. in diameter, 5 mm. *above* d.a.; Camescasse[76] (1885), a thin diaphragm at ductus almost obliterated lumen; Hale-White[77] (1884); Barié[78] (1886); Alexais and Gilly[88] (1887); Loriga[89] (1887); Flint[93] (1895); Kretz[92] (1895); Wadstein[94] (1897), Case 2;[95] Case 3‡;[144] Schichhold[96] (1897); Smith and Targett*[143] (1897), a sudden narrowing at obliterated ductus as though by a septum with central oval aperture 3.16 inch in greatest diameter; MacCallum[105] (1900); Bumke[145] (1901), 4 mm. long annular constriction admitting a 1 mm. sound exactly at insertion of ligamentum arteriosum, aorta here kinked and spirally twisted; Reinitz*[146] (1902), Case 1, sudden constriction as though by a cord admitting a match; Case 2,*[147] a knitting needle; Libman[99] (1902); Pic and Bonnamour[100] (1902); Bonnet[101] (1903), sudden constriction as though by a ligature deeply grooved on convexity, just below obliterated ductus internally, culdesac formed by circular diaphragm 1 mm. thick with central opening admitting a fine sound and expending widely below; Wasastjerna†[106] (1903), constriction below d.a. admitting fine sound; Murray*[149] (1904); Fawcett† (1905); Cases 2[107] and 11;[111] Pappenheimer,† Case 1[117] (1906); Case 2;[118] Mackenzie†[121] (1908); Monckeberg†[119] (1907), Case 1; Case 2;[120] Sella,† Case 1[123] (1910); Oberndorfer†[125] (1910); Borissowa*[153] (1910); Fraenckel,* Case 2[156] (1912), rapid narrowing of aortic lumen beginning at carotid and reduced to 1 mm. before origin of left subclavian; Erdmenger*[154] (1912); Kolisko,* Case 1[159] (1913) only very thin metal sound could pass aorta at obliterated ductus; Berger*[158] (1913); Weidman*[162] (1914); West†[128] (1919); Hinrichsmeyer‡[166] (1919), funnel-shaped constriction admitting sound at d.a.; Hart,* Case 2[168] (1920); Strassman* (1922), Cases 4;[170] 6;[172] 7;[173] Katz*[169] (1921); Follet and Caille†[129] (1921); Beneke*[176] (1922), just below ductus aorta diameter of thin pencil, and above this low comb-shaped diaphragm of thickened intima; Focken† (1924), Cases 1[132] and 2;[133] Nieuwejaar‡[178] (1925), marked narrowing of otherwise apparently healthy aorta as though ligatured below obliterated ductus, with diaphragm internally, leaving lumen 1.5 cm. in circumference; Mackenzie*[181] (1927), aortic wall constricted and a thin fold projects into lumen, leaving orifice 3 mm. wide and 4.5 mm. high in line with closed ductus; Parkes Weber*[183] (1927), a typical abrupt diaphragm-like stenosis below left subclavian just admitting a probe 2 mm. in diameter; and the following from Blackford's additional cases: Bartels§;[194] Hansteen§, Case 2;[198] Lutzow-Holm§;[200] Santas§;[191] Maixner§;[186] Kovesi§;[190] Lantinga§;[185] Turnbull§;[195] Broome§;[188] Josefson§.[189]

There are thus among these 200 cases with autopsy in adults here reviewed 155 cases of grave coarctation in which the arch of the aorta was either completely obliterated (in 47 cases), or was so narrowed as not to exceed the diameter of a crowquill (6 mm.) (in 108 cases). In the remaining 45 cases the stenosis was of a "moderate" grade, admitting the forefinger, "about one-third of its normal caliber," etc.

*New series in this and Blackford's article.
†Series in Abbott's monograph, Osler's System.
§New series in Blackford's article.
‡New series, this article only.

There are, as indicated above, many other such slight cases of coarctation in adults in the literature, especially complicating other defects, but these 45 are included here, not only because most of them have been published as such, or listed in other series, but also because even these mild degrees of stenosis may apparently have a practical bearing on the duration of life, as is shown in the cases reported by Sir Thomas Horder,[151] Binder,[165] and Bronson and Sutherland,[163] in which death occurred from spontaneous rupture of the aorta; as also in the frequent combination of a bicuspid aortic valve which so often acts as a nidus for bacterial infection and resultant cardiac disease.

Following is a list of the 49 45 cases of *moderate stenosis* in this series:

Sir Astley Cooper[3] (1818), stricture at d.a. formed by thickening of fibrous coats admitting with difficulty the little finger; Laennec[59] (1826), a hollow admitting almond at attachment of ligamentous d.a. with septum at upper edge of this formed of all coats of the vessel; Nixon[8] (1834), aorta reduced to half its caliber; Maigne[60] (1837), double stenosis above and just below left subclavian, lower barely admits finger; Fletcher[16] (1842), caliber diminished one-third at d.a.; Chevers[21] (1845); Crisp, Case 1[62] (1847), stenosis at level of d.a.; Dubreuil, case 1[79] (1847), reduced to ¼ normal diameter at ligamentous ductus; Babington†[102] (1847); Blakiston, Case 1[23] (1848), just beyond left subclavian aortic wall thinned and diameter greatly narrowed; Case 2,[24] at d.a. aortic coats thickened and caliber reduced one-half for about an inch; Willigk[41] (1853); Schmidt[42] (1853), stenosis at d.a. scarcely admits finger, coats thickened; Härlin[32] (1855), double stenosis, lower at level of d.a. barely admits tip of little finger; Peacock†[103] (1862), infantile type; Church†[104] (1865), narrowing to diameter of 1 inch; Quinquand[66] (1868), below left subclavian stenosis for 7 cm. barely admitting end of index finger; Purser[55] (1873); Oehl‡[139] (1875); Moore, Case 2[84] (1876), sharp constriction just admitting index finger; Kriegk, Case 3[58] (1878), annular, at d.a. admitting little finger; Lesseliers*[142] (1882); Fawcett† (1905), Cases 5, 6, 12, 13, 14, 15, 16[109-116]; Umber*[150] (1905), aorta exquisitely hypoplasic, narrowed to 4 cm. in circumference at obliterated d.a.; Horder*[151] (1907), slight narrowing at patent d.a.; Kohn*[152] (1911); Stauing*[157] (1913); Bronson and Sutherland*[163] (1918), aortic isthmus 0.7 cm. (infantile type); Binder*[165, 119] isthmus 3.5 cm. in circumference, slight further contraction at (closed) d.a.; Goldblatt†[131] (1922), lumen reduced at d.a. by cicatricial ridge; Reifenstein†[134] (1924); Laubry*[179] (1926); Smith and Hausmann†[135] (1926), aorta constricted to half normal size at site of (closed) d.a.; Libman‡[180] (1927); and from Blackford's series: Kurcyuoza§[184] (1874); Edgren§[187] (1897); Santas§[191] (1905); Hansteen, Case 1§[197] (1925); Case 3§.[199]

AGE INCIDENCE OF "INFANTILE" AND "ADULT" TYPE OF COARCTATION

The brief description already given of the more important variations among the 200 cases of coarctation in adults here analyzed shows that a relatively small percentage of the cases actually do show a continuance into "adult" life of the "infantile" type, in that the abnormality consists in a simple narrowing or persistence of the fetal isth-

*New series in this and Blackford's article.
†Series in Abbott's monograph, Osler's System.
‡New series, this article only.
§New series in Blackford's article.

mus. On account of the fact that it is not always possible from the frequently inadequate descriptions of published cases to decide upon the exact anatomical structure of the stenosis, it has been found necessary, as shown in the title, to limit this study to the obstruction in the descending arch in subjects over two years of age (i.e., adults). This necessarily arbitrary division unfortunately makes it necessary to exclude from this series those rare cases in which the adult type of coarctation has been found to be present at or shortly after birth, as in those reported by Ettlinger[232] of an infant seventeen days old, and by Wadstein[233] of an infant seven months old. Both these were classic examples of the adult type in that the aorta was (a) partly, or (b) entirely occluded by an intimal shelf or diaphragm which projected into its lumen at or immediately below the insertion of the ductus. These and other observations in which Bonnet's "adult type" has actually been demonstrated to exist at or shortly after birth, are very important from the standpoint of etiology and may not be lost sight of, although the cases are perforce not incorporated in the present series.

ASSOCIATED ANOMALIES

A great deal of interest attaches to the presence or absence of other anomalies and the form of those which are associated. As Bonnet[101] first pointed out, this is an important differential point, in that the graver and more complex anomalies, such as biloculate or triloculate heart, transposition of the arterial trunks, pulmonary atresia, etc., are very commonly combined with the "infantile" type but are practically never found associated with the *extreme degrees* of the classic adult form; while on the other hand, what may be called "minor" anomalies, such as bicuspid aortic valve, anomalous origin of the arteries from the arch, persistent left superior vena cava, defects of the aortic septum, and subaortic stenosis occur relatively so frequently in the adult type as actually to appear to form part of a significant anatomical complex. The contrast is well seen in the analysis of 212 cases of both types which I made in another connection.[204] Among 82 of these which were in "infants under one year," 50 were complicated by "grave" or "major" anomalies, and 19 by "minor" ones, while in only 15 cases was the defect uncomplicated by any other anomaly. On the other hand in the 155 of these cases in which the coarctation was in subjects *over* one year ("adults"), there were only 13 cases complicating "grave" anomalies, against 57 in which "minor" anomalies were associated, and 86 in which the coarctation was the only defect. In these 13 cases complicated by other "grave" anomalies,* the coarctation was either so slight as to be of

*These 12 cases of coarctation in "adults" in grave associated anomalies are by: Grunmach (dextrocardia with septal defect) Berl. klin. Wchnschr., 1890, xxvii, 22;

no apparent importance while the other anomaly dominated the clinical picture, or it was of the infantile type, the pulmonary artery forming the aorta through a widely patent ductus (Mackenzie, Habershon, Chiari). For these reasons these 12 cases have been excluded from the 200 which form the basis of this article, as have also the following 18 cases in which a slight degree of coarctation complicated "minor" anomalies which were classed as the "primary lesion in my monograph. The latter are of special interest because, as mentioned above, these particular forms of defect are relatively so frequent in the adult type of coarctation that a common or like error in development may well have initiated both anomalies. On this account they are enumerated as follows:

Luksch: (M. aged 76 years, anomalous chorda, in aorta; ascending aorta 8 cm., isthmus 5 cm. in circumference.) *Centralbl. f. allg. Path. u. path. Anat.*, 1912, xxiii, 626.

Abbott: (F. aged 28 yr., large patent foramen ovale, hypoplasia of aorta, 4 cm. at origin, 3 cm. below left subclavian.) *Bull. Int. Assn. Med. Mus.*, 1915, v, 129.

Cramer and Frommel: (F. aged 41 yr. Defect 3.5 by 2 cm. in auricular septum with mitral stenosis, aorta hypoplastic with slight coarctation at insertion of obliterated d.a.) *Arch. d. mal. du coeur*, 1923, xvi, 560.

Greenfield: (M. aged 53 yr. Defect upper part auricular septum, all pulmonary veins enter right auricle.) *Jour. Anat. and Physiol.*, 1890, xxiv, 423.

Sternberg: (M. aged 16 yr., large defect of A. S. below, cleavage of anterior mitral segment.) *Verhandl. d. deutsch. path. Gesellsch.*, 1913, xvi, 253.

Kraus: (M. aged 27 yr., congenital aneurysm right aortic sinus of Valsalva.) *Berl. klin. Wchnschr.*, 1902, xxxix, 1161 (Case 1).

Thursfield and Scott: (M. aged 14 yr., subaortic stenosis and thin line of fibrosis in otherwise healthy aorta just above aortic cusps.) *Brit. Jour. Dis. Child.*, 1913, x, 104.

and the following cases complicating a widely patent ductus arteriosus.

Hochhaus: (M. aged 24 yr. Slight ridge above patent d.a.) *Deutsch. Arch. f. klin. Med.*, 1893, li, 1.

Hamilton and Abbott: (F. aged 19 yr.) *Tr. Assn. Am. Phys.*, 1914, xxix, 294.

Stoddard: (F. aged 17 yr.) *Arch. Int. Med.*, 1915, xvi, 38.

Wells: (M. aged 42 yr.) *Am. Jour. Med. Sc.*, 1908, cxxxvi, 381.

Greenhow: (F. aged 17 yr.) *Tr. Clin. Soc., London*, 1876, ix, 152.

Luys: (F. aged 58 yr. Pulmonary artery forms descending aorta through dilated patent d.a.) *Mém. Soc. de biol.*, 1856, p. 74.

Schrötter: (F. aged 5 yr.) *Ztschr. f. klin. Med.*, 1901, xliii, 161.

Duroziez: (M. aged 40 yr.) *Mém. Soc. de biol.*, 1862, 3rd series, iv, 285.

Mackenzie (heart with three ventricles) Tr. Path. Soc. London, 1880, xxxi, 63; Libman (dextroposition of aorta with ventricular septal defect) Rept. by Abbott, Ann. Clin. Med., 1925, iv, 197; Cockle, Med. Chir. Trans., xlvi, 193; Meinertz, Virch. Arch., clxvi, 385, (transposition of great trunks); Finlay, Tr. Path. Soc., London, 1879, xxx, 263; Lafitte, Bull. Soc. Anat. de Paris, 1892, vi, 13; King, Tr. Path. Soc., 1872, xxiii, 83; Variot, Jour. de clin. et de thérap. inf., 1897, p. 381; Moore, St. Barth. Hosp. Rep., 1875, i, 225 (pulmonary stenosis); Habershon (pulmonary atresia) Tr. Path. Soc. London, 1888, 71 (enumerated among the 155 cases in the writer's monograph); Chiari, Jahrb. f. Kinderh., 1879, p. 219 (from Bonnet's series); and Houel, Manuel d'anat. path. de Paris, 1879 (from Barié's series); and to these may be added a fourteenth case, that by Kurtz, Sprague and White§ (pulmonary stenosis with ventricular septal defect).

Wasastjerna: Case 1. (M. aged 13 yr.) *Finska läk-sallsk. handl.*, 1874, xvi, 235.
Wasastjerna: Case 2. (M. aged 36 yr.) *Schmidt's Jahr.*, 1876, clxix, 142.
Bonner: Case 1. *Schmidt's Jahrb.*, clxiv, 142.

To the above Dr. Blackford adds 2 cases, 1 of *subaortic stenosis* (Langwill, *Scottish Med. and Surg. Jour.*, 1897, i, 723), and 1 of aortic stenosis and hypoplasia (Anders, *Med. News*, 1902, liii, 827), making in all 20 cases complicating minor, in addition to the 14 cases, enumerated above, complicating major anomalies. This adds to the 200 well-pronounced cases of coarctation in adults from the literature in this series 34 more cases of stenosis of a moderate grade or of infantile type complicating other anomalies classed for reasons given above as the "primary lesion."

Turning now to the incidence of other defects in the 200 cases (from which, following Bonnet, the slighter degrees of coarctation complicating grave abnormalities of the cyanotic group have been excluded), we find among these a highly suggestive set of "minor" associated anomalies, which duplicate in some instances those mentioned above as having been combined with a mild grade of stenosis of the descending arch not included in this series. A careful revision of the available data concerning the 200 cases of this series yields the following list, which would undoubtedly be larger if all the original records could have been examined. From it, it is evident that by far the most frequent anomalies associated with coarctation in adults are (a) an abnormal origin of the arteries from the arch or their branches, or (b) a bicuspid state of the aortic valve. These are also of the greatest etiological significance and clinical import of all associated anomalies. A third condition of importance from the latter standpoint is (c) subaortic stenosis. These subjects will be further discussed below.

Associated Anomalies of the Body at Large—Somatic Defects.—The following small group forms an interesting link in the chain of evidence pointing to developmental arrest as the chief causative factor in the adult as well as in the infantile form:

Hypospadias (Wood[37]); absence of left kidney, ureter and vesiculae seminales (Reinitz[146]); vertical position of stomach, embryonic position (Martens[90]); horseshoe kidney (Broome§[188]); diaphragmatic hernia, subluxation of joints, slight mongolianism (Bronson and Sutherland[163]). The two last named were of the infantile type, the first three were classic examples of the adult form. Ichthyosis (Church[104]); pigeon-breast (Sella's Case 2;[123] Taruffi;[140] Beneke[176]).

Associated Anomalies in the Heart Itself.—

Large defect at *lower* part of auricular septum, persistent ostium primum, with cleavage of anterior mitral segment and funnel-shaped mitral orifice (Strassner[122]); large defect at *upper* part of interauricular septum above patent foramen ovale, and displacement of all pulmonary veins into right auricle (Fawcett,[112] Obs. 12); widely patent foramen ovale (Kriegk,[58] Church,[104] Kohn,[152] Fraenckel,[156] Case 2); perfora-

§Blackford's Series.

tion between left auricle and aorta, (?) congenital (Loriga[89]); localized defect at base of interventricular septum (Fletcher[16]); aneurysm of pars membranacea (Martens[90]); with deformity of mitral orifice (Wilks[36]); *diverticulum just above aortic cusp into right auricle* (Flint[93]); *subaortic stenosis* (Rokitansky's Obs. 2030; Taruffi[140]; O'Flaherty[50]; Hamilton and Abbott[136]); anomalous chorda in left ventricle (Eppinger[48]); dextroversio cordis (Bahn-Pol[177]).

Here are 18 cardiac anomalies among the 200, all apparently produced by some slight arrest or derangement of growth late in embryonic life, i.e., just before or at the time of closure of the cardiac or arterial septa. In 6 cases this was of the nature of an auricular defect, and there are 4 other such, mentioned above, among the slighter grades of coarctation not included in this series; of these ten, two (one in this series by Strassner[122] and one not included by Greenfield [see above]) are cases of *persistent ostium primum,* while in two others the defect lay at the extreme upper border of the auricular septum and all the pulmonary veins emptied into the left auricle. Similarly there are 4 cases in this series in which *subaortic stenosis* complicated the coarctation, and to these we may add the two other well-marked examples of this condition complicated by lesser grades of coarctation, reported by Thursfield and Scott and Langwill. Finally, in the two sets of cases there are 2 examples (Kraus[234] and Flint[93]) of that rare anomaly known as congenital aneurysm of the aortic sinus of Valsalva, which finds its homologue in the foramen panizzae of the crocodile and is certainly to be explained by an arrest of growth at the end of the critical period of cardiac evolution.

Persistent Left Superior Vena Cava.—This occurred in three cases, all of the classic adult type, in which the aorta was almost obliterated by a constriction resembling a ligature at or below the insertion of the ductus arteriosus (Bonnet,[101] Kolisko,[159] Broome§[188]). Here again we meet with an anomaly which is very characteristic of an arrest in late embryonic life, and one that links the adult with the infantile type as being dependent upon a like or common cause; for persistent left duct of Cuvier is a very common happening in stenosis of the isthmus in the newborn.

Anomalies of the Arteries Taking Origin From the Aortic Arch.—Here follows an exceedingly interesting and highly suggestive series:

Accessory right coronary (Fraenckel,[156] Hamilton and Abbott[136]); deep cervical from left subclavian artery (Meckel[5]); both inferior thyroids from common carotids (Hale-White[77]); anomalous branch from basilar artery enters cavernous sinus, falx cerebri deficient (Fawcett[115]); both carotids from a common trunk, vestigial remains of double ductus arteriosus (Hamernjk's Case 1[20]); innominate and left carotid from common trunk (Maigne,[60] Murray[149]); left vertebral from arch (4 cases, by Bradley,[82] Legg,[85] Haberer,[148] Kolisko[159]); first part of left subclavian a fibrous cord, ending in bulbous lower end of pervious second part (Woltman and Shelden[182]); right subclavian arises from descending arch below left and passes

§Blackford's Series.

obliquely up behind trachea and esophagus (Hamernjk, Case 3[40] and Fawcett[112]); anomalous artery from narrowed isthmus immediately above point of obliteration, (?) persistent fifth left arch (Hamilton and Abbott, this case[136]); supernumerary branches above constriction entering aorta below this (Wasastjerna[106]); accessory right aortic intercostal (Reynaud,[6] see Fig. 9); aneurysms of cerebral arteries, (?) congenital (Eppinger[49]; Kolisko[160]; Strassman[173]; Woltman and Shelden[182]; Parkes Weber[183]).

The significance of certain of the above cases, especially such as present an anomalous origin of the great trunks, becomes immediately apparent when one compares Rathke's diagrams of the primitive aortic arches and the involution which certain of these undergo at successive stages of embryonic development. Assuming that the explanation of such classic cases of the adult type of coarctation as that which forms the subject of Part I of this article is an atrophy of the descending part of the left fourth arch at or near its junction with the left sixth (ductus), it is evident that irregularities are liable to occur in the contiguous parts of the other primitive arches, or even in the structure of the whole vascular *anlage*, as a result of the action of the same cause, so that the presence of the associated anomaly is of material assistance in fixing the approximate date of origin of the coarctation. In this connection the diagrams by Dr. H. A. Harris[235] (Fig. 5) illustrating the various stages in the involution of those embryonic arches that are predestined to disappear in man, as mirrored phylogenetically in the piscian, amphibian, and reptilian heart, are highly illuminating. Thus, the curious phenomenon of the right subclavian arising from the descending aorta below the left (Hamernjk, Fawcett) is to be explained as an atrophy and disappearance of the *proximal* part of the right fourth arch (which normally becomes the right subclavian) and the persistence of its *distal* portion, which normally becomes obliterated in man, although in birds and reptiles it persists as the lower part of the right (in birds), or double, arch of the aorta. Similarly, in Woltman and Shelden's case the atrophy of the first part of the subclavian artery is merely an extension of the same process that has involved the immediately contiguous descending part of the right fourth arch with which it is directly continuous and which is the seat of the coarctation. Again, the origin (in 3 cases) of the left vertebral from the arch instead of from the left subclavian, would appear to be due to the taking up of the beginning of the latter in the wall of the aorta in the obliterative process that is occurring below; while the persistence of the left fifth arch, which is a unique feature of our case, is apparently incidental to the same process of arrested development.

In another group of these cases the anomaly appears to consist in the actual development of new branches from the aorta to assist in the collateral circulation. This was apparently so in Wasastjerna's

remarkable case in which four supernumerary arteries rising just above the constriction appeared to communicate with four other anomalous branches from the aorta just below this, thus making a short circuit for the deflected blood, as also in Schlesinger's[236] atypical case of obliteration of the lower thoracic aorta, in which "many un-

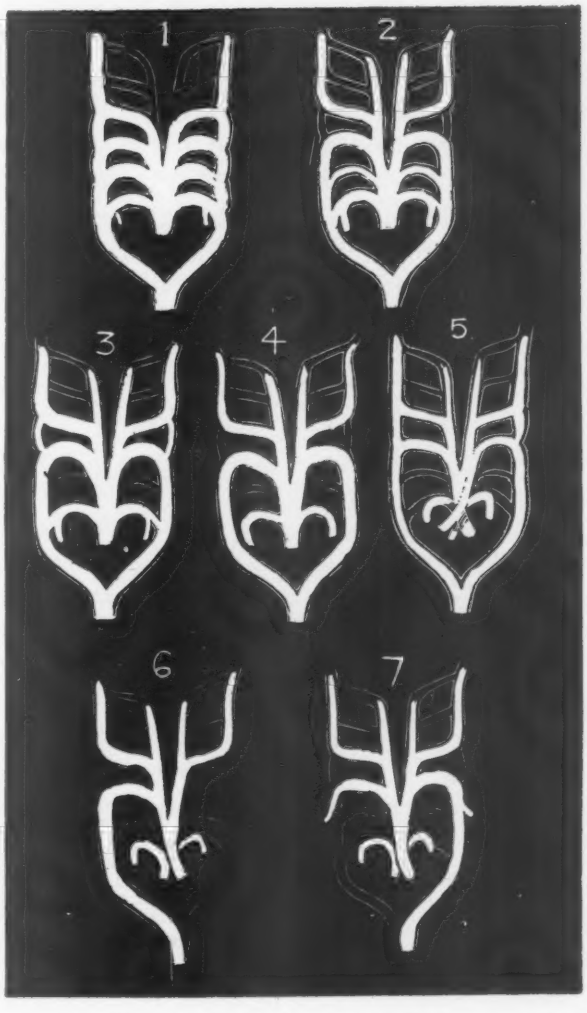

Fig 5.—The aortic arches and their transformations (after Rathke). *1*, ceratodus; *2*, salamander; *3*, triton; *4*, frog; *5*, lizard; *6*, bird; 7, mammal. The critical point is the junction of the sixth left arch with the descending thoracic aorta. (From the article by H. A. Harris, *Jour. of Anat.*, 1922, lvii, 85, Fig. 5.)

usual branches from the descending aorta united with the internal mammaries and subclavians."

The cerebral aneurysms, multiple or simple, which have been demonstrated in some of the most extreme cases of adult coarctation are believed by the general consensus of opinion to be congenital. This subject will be discussed further.

Bicuspid Aortic Valve and Other Anomalies of the Semilunar Cusps.—Discussion of the remarkable frequency of congenital abnormalities of the semilunar cusps and, especially of the bicuspid aortic valve in the adult type of coarctation, is quite beyond the range of coincidence. Reinitz[146] placed the incidence of the latter at 10 per cent, but this is certainly far too low. In the 183 cases accessible to us there were 46 cases (that is 25.1 per cent) in which the aortic valve was apparently congenitally bicuspid, and 4 others in which the description and the presence of inflammatory changes did not permit one to exclude an acquired fusion, although even in these their association with typical cases of adult coarctation argued for their congenital origin (cases of Goodhart,[71] Barth,[28] Staunig,[157] Parkes Weber[183]). In 6 other cases among the 183 listed below, miscellaneous anomalies, such as supernumerary or defective aortic segments, existed.

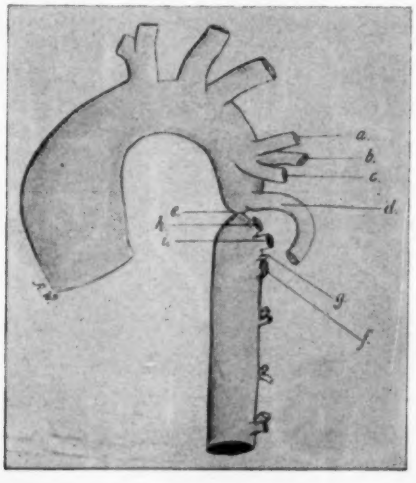

Fig. 6.—Aortic arch from Wasastjerna's case of extreme coarctation showing four anomalous branches arising from the descending aorta immediately above (*a, b, c, d*) and below (*f, g, h, i*) the constriction (*e*), which admitted a fine sound 1 mm. thick. The opening (*f*) at the lateral border of the descending thoracic aorta apparently connected with the horseshoe-shaped vessel (*d*) just above the constriction, but was cut at the autopsy. Just at the stenosis (*e*) was a scarred area indicating the site of the ductus. In a healthy boy aged thirteen, dying of rupture of the aorta. (From article by E. Wasastjerna, *Ztschr. f. klin, Med.*, 1903, xlix, 405.)

In considering the incidence of bicuspid aortic valve in coarctation, it has been necessary to differentiate, in these reported cases, between a congenital lesion and an acquired fusion, from the description of the gross appearances alone. Where the existence of only two cusps, often of large size, without any trace of a third is described, as was the case in 28 of this series listed below, an antenatal arrest is plainly manifest; but where, as in the remainder of these 50 cases the presence of a third segment was indicated by a more or less well-marked raphe or partition, indicating the junction of the combined segments, the problem is less simple, especially as the condition is nearly always obscured by superimposed inflammatory changes. The criteria of a

congenital origin, first laid down by Sir William Osler[237] in his classic communication on this subject before the Association of American Physicians in 1886, supply the necessary distinctive features. These are (a) the presence of a low, almost obliterated raphe incompletely dividing the sinus behind the composite cusp; (b) compensatory changes, which are evidently prenatal, in the relative length of the two segments, the single one frequently becoming the larger, and the composite cusp shorter than or equal to the free border of the former by direct measurement; (c) the unchanged condition of the ventricular surface of the combined segment in absence of puckering or such tissue changes about the commissure as would be bound to result from postnatal fusion; (d) absence of the nodula Arantii of the normal cusp, as occurred in all Osler's 18 cases of bicuspid aortic valve and is mentioned in several of our cases; (e) the occasional absence of all trace of inflammatory thickening in the combined cusp; (f) changes in the attachment of the composite cusp to the aorta; (g) the relative frequency with which the coronary segments are fused. To these Parkes Weber added the observation that the form of the raphe, which in one of his cases was fenestrated, may give proof of its congenital origin (as in my own case). These early deductions from observation of the gross appearances have been confirmed and the distinction between the congenital and acquired lesion established histologically by the recent epoch-making studies of Sir Thomas Lewis and Dr. R. T. Grant[238] upon the architecture of the aortic valve in so-called congenital and in acquired fusion, in which the low raphe seen behind the conjoint segment was shown by reconstruction of serial sections to be not an incomplete fusion of two originally independent segments, such as might have been produced by a fetal or very early postnatal endocarditis, but a true malformation, an arrest of growth of the annulus fibrosis and covering layers of the valve, so that the several normal layers of one cusp pass uninterruptedly into the other across the region normally broken by the commissure. These workers also showed statistically that the favorite combination is between the coronary cusps, both in the congenital lesion and in acquired fusion, and that the commissure between the posterior and left cusps is rarely involved.

In the following descriptive list of the *bicuspid aortic valves* in this series, the cases are grouped according to the (a) absence, or (b) presence of evidence of a third segment, and those cases in which the coarctation took the form of obliteration or extreme stenosis of the descending arch are in italics. Termination by rupture of the aorta or cerebral hemorrhage is also indicated.

(a) *Only two cusps:* Running from back to front and parellel (Otto,[4] rupture of aorta); one large and one small cusp which formed a funnel projecting into the aorta (Blakiston[23]); one anterior and one posterior (*Hamernjk*[58]); only two aortic

cusps, well formed in other respects (*Kriegk*,[60] cerebral hemorrhage); only two aortic cusps, thickened, ossified, and insufficient (*Moutard-Martin*[70]); aortic orifice presented only two aortic cusps and a well-marked insufficiency (*Liouville*[81]); by a congenital anomaly, the aortic cusps are reduced to two, posterior and anterior, the latter rigid and immobile with calcareous plate (*Alexais and Gilly*[88]); only two aortic cusps, anterior and posterior, indurated and insufficient to the water test (*Dickinson and Fenton*[98]); only two aortic cusps, sinuses of Valsalva very deep, rupture of ascending aorta (Wadstein‡[144]); aortic orifice wide, has only two segments (Fawcett†[115]); anterior segment absent, great hypertrophy of the other two with thickening (Mackenzie†[121]); aortic valve bicuspid and sclerosed, no sign of raphe (Monckeberg†[119]); only two cusps without raphe, no noduli Arantii, rupture of ascending aorta (*Fraenckel**[156]); only two cusps, one long, one short, rupture of ascending aorta (Binder*[165]); two large cusps, not fused, thick at edges with verrucose endocarditis (*Hart**[167]); only two aortic cusps, both thin and healthy, no sign of raphe, rupture of ascending aorta (Goldblatt†[131]); only two cusps, one anterior, one posterior, rupture of ascending aorta (*Nieuwejaar*‡[178]); only two cusps (rupture of cerebral aneurysm, *Eppinger*[49]); (*Legg*[85]); (*Pappenheimer*†[117]); (with rupture of ascending aorta, *West*,*[137] *Strassman**[172]); aortic valve bicuspid (rupture of mycotic aneurysm Smith and Hausmann†[135]), (mycotic aneurysm *Focken*†[132]); (Laubry*[179]); (*Weidman**[162]); (*Umber**[150]); only two perfect cusps, (*Woltman and Shelden*,*[182] rupture of cerebral aneurysm).

(b) *Bicuspid with raphe:* Two cusps, slight raphe behind one (Wilks[36]); anterior and left cusps blended to form a single curtain 2½ inches long with low raphe behind it, right cusp 2¼ inches long, no corpus Arantii, rupture of aorta ascendens (*Barker*[39]); aortic cusps half as large again as normal, right and left fused and separated from wall at their common commissure, posterior (single) cusp twice as long as anterior, rupture of ascending aorta (*Hornung*[74]); two cusps with low raphe behind, thickened and insufficient (*Traube*[51]); only two aortic cusps, left much enlarged, the right formed from fusion of right and posterior segments, both elongated, close but narrow aortic orifice, cerebral hemorrhage (*Sommerbrodt*[75]); only two aortic cusps, left and right fused into a single one (*Loriga*[89]); by a congenital anomaly right and posterior aortic cusps are fused to form a single segment, ulcerative endocarditis of this (*Wadstein*[95]); posterior cusp broad and thickened, left broad and fused with right, free border rounded, rupture of ascending aorta (*Sella*†[123]); right and left cusps fused, low raphe behind one, rupture of ascending aorta (*Sella*†[124]); cusps thickened and two fused, rupture of ascending aorta (*Oberndorfer*†[125]); with congenital fenestrated raphe, cerebral death, impending rupture of aorta (*Hamilton and Abbott*†[136]); right and posterior cusps fused but not thickened, healthy and competent, rupture of ascending aorta (Bumke*[145]); only two cusps with raphe, cerebral hemorrhage (Reinitz,*[146]; Erdmenger*[154]); two very broad cusps, anterior and posterior, coronaries separated by low raphe behind anterior rupture of ascending aorta (*Meixner**[175]); cusps fused with low raphe (Haberer*[148]); two cusps fused thin and healthy (Taruffi‡[140]); two cusps with low raphe (Hinrichsmeyer‡[166]).

In 4 other cases the description given did not preclude a postnatal origin. These were as follows:

Coronary cusps fused and thickened (*Barth*[28]); two sigmoids fused, at their point of junction a hard fibrinous mass (*Goodhart*[71]); fusion of anterior aortic cusps, rupture of ascending aorta (Staunig*[157]); anterior segments fused and thickened, ruptured cerebral aneurysm (*Parkes Weber**[183]).

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*New series in this and Blackford's article.
†Cases in Abbott's monograph, Osler's System.
‡New series this article only.

making 50 cases among the 183 accessible to us, in which the aortic valve was bicuspid, either from congenital or from postnatal causes. To these may be added Maixner's[186] case of congenitally bicuspid valve from Dr. Blackford's own series, making, among the entire 200 cases, 51 of bicuspid aortic valve, of which 47 have been concluded, from the data given, to be actually congenital.

Miscellaneous Anomalies of Semilunar Cusps Other Than Diminution in Number.—Six cases, which is only 1.3 per cent of the total anomalies of these cusps in the 183 cases studied.

Accessory cusp (Wilks[36]); accessory (fourth) cusp in aorta ¼ inch above valve (Babington†[102]); multiple fenestrations (*Monckeberg*†[119]); two large and one small aortic segment, malignant endocarditis, and mycotic aneurysm at coarctation (*Focken*†[132]); one aortic segment abnormally small (Lesseliers*[142]); three healthy cusps, the right very big and so approximating to left that the two look like a single segment, ascending aorta very thin and ruptured spontaneously (*Berger**[158]).

It is scarcely necessary to remark that the presence of anomalies of the semilunar cusps and especially of the bicuspid aortic valve in the adult type of coarctation is not merely of academic interest, as indicating in a very striking manner the antenatal origin of this anomaly in later embryonic life. From the practical clinical standpoint, this combination is of the most serious import, and is significant of all the great dangers to which the patient stands exposed, for, in the first place, a bicuspid aortic valve directly lowers his expectation of life by presenting a *locus minoris resistentiae* for the invasion of infections, thus exposing him to the immediate and remote effects of cardiac complications, both aortic valvular disease and a damaged myocardium, as well as the more severe bacterial inflammations which find a special nidus for localization in crevices of the composite cusp ("commisural lesion" of Lewis and Grant[238] or "chronic infectious endocarditis" of Osler[239]), and which practically always terminate fatally, and may serve also as the initial focus for a mycotic aneurysm; while the chronic valvular and myocardial lesions that result from the milder grades of infection combine to lower the cardiac reserve and render this inadequate for the demands of the collateral circulation, decompensation following.

In the second place (as is manifest from the preceding list) the presence of a bicuspid aortic valve appears to indicate, at least in a portion of the cases in which it occurs, a tendency to *spontaneous rupture of the aorta,* which hangs always, like a Sword of Damocles, above the unsuspecting subjects of this type of coarctation, for this anomaly occurred in quite half the cases so terminating. A causal relation between the three conditions is not clear, but it seems probable

*New series in this and Blackford's article.
†Cases in Abbott's monograph, Osler's System.

that the thinning, which is not infrequently seen in the wall of the ascending aorta in these cases, may be also of congenital origin and due to the same arrest of development that led to an incomplete evolution of the endocardial cushions destined to become the aortic cusps. That such a thinned area would yield the more readily to form a dissecting aneurysm, with later rupture under the increased pressure that exists in the upper part of the body in adult coarctation, would appear to be self-evident. Extension of such a congenital weakening of the media to the whole arterial system might similarly explain the coincidence of cerebral hemorrhage and such a process might even have a part in the peculiar changes at the area of coarctation itself.

Congenital Hypoplasia of the Aorta.—In a relatively small proportion of the cases of coarctation in adults, a hypoplasia of the arterial system as a whole occurs. This is evidenced by a narrow ascending aorta and aortic arch with elastic and usually thin walls, and by the slender build, gracile delicate frame, and delayed development of the subject, who may present the characteristic clinical picture of a generalized hypoplasia of the aorta with the features superadded of an obstruction in the descending arch. Owing to the fact that the ascending aorta is so commonly secondarily dilated as a result of the coarctation, the original narrowing at this point may be obscured in patients attaining adult life, so that the association of a congenital hypoplasia of the arterial tract may be more frequent than is apparent from the reports; and conversely, the post-mortem report is not always sufficiently detailed to permit of a positive conclusion as to the presence or absence of dilatation. There are among the 200 cases, however, 21 (i.e., 10 per cent) in which the ascending aorta was definitely narrower than normal, and in which this may, from the context, be considered to be of congenital origin. These are the following:

Barth,[28] aorta diminished in ascending portion; Kriegk,[58] aorta at origin is 5.4 cm. in circumference; Andral[61] (male, aged 22) whole aorta including the ascending portion was very hypoplastic, barely admitting the index finger; *Almagro*[65]; *Riegel*[68] (male, aged 29) aorta at origin 0.055 mm. in diameter, hypoplasia of whole arterial system; *Martens*[90] (male, aged 24) aorta at origin 50 mm. in circumference, near innominate 40 mm. *Wadstein*[95] (female, aged 22) aorta everywhere narrow and thin-walled; Babington,†[102] female, aged 34, aorta 2 1/10 inches at origin, narrow throughout; Peacock,†[103] aorta very small, diminishes further after left subclavian; *Fawcett*,†[111] aorta smallish and strong; Fawcett,†[113] aorta was narrow and its valves were thin; *Monckeberg*†[120] (female, aged 26) ascending aorta narrow, smooth and elastic, 4.8 cm. in circumference at origin; *Focken*,†[132] aorta smooth and elastic, 6 cm. in circumference; Umber*[150] (male, aged 22) pale, delicate frame, aorta exquisitely hypoplasic throughout; *Borissowa*,*[153] aorta at origin only permeable to a sound; *Erdmenger**[154] (male, aged 24) of medium size, gracile bony framework, marked pallor of surface, weak

†Series in Abbott's monograph, Osler's System.

musculature, aorta at origin 6.5 cm. in circumference; *Weidman*,*[162] ascending aorta 2.5 in circumference; Taruffi‡[140] (male, aged 19) of puerile habitus and genitalia, ascending aorta smooth walled, not enlarged; also, from Blackford's series, Hansteen's Case 3,§[199] Kurcyuoza§[184] and Lantinga,§[185] all of generalized hypoplasia.

SECONDARY CHANGES

Dilatation of the Ascending Aorta.—In the great majority of the cases of adult coarctation the ascending aorta immediately above the aortic ring, and to a greater or less extent the aortic arch just above this, is definitely dilated. This was so in 101 of the 200 cases before us (50 per cent), and this figure is certainly below the actual proportion, for in 71 others this point was not mentioned, while in 21 cases the aorta was hypoplasic, and in only 7 was it said to be normal. The increase of the lumen varies from a slight enlargement (8 to 9 cm. in circumference) to a great widening or ballooning of the wall (most commonly of its left posterior aspect) 13 to 14 cm. in circumference, which commonly passes on to a diffuse or saccular aneurysm, or may become the seat of a dissecting aneurysm, and this in turn may rupture into the pericardium or adjacent viscus. The cause of the dilatation of the aorta in this situation is no doubt in part the increased intravascular tension that exists in the upper part of the body above the stenosis, under which the great branches of the arch also become dilated and often atheromatous; but it is also undoubtedly due in large part to an inherent weakness of the middle coat of the aortic wall itself, which is evidenced in some cases by this being "as thin as parchment" (West,*[137] Sella,†[124] Meixner,*[175] Berger*[158]), and by the frequency with which dissecting aneurysms occur. In this connection the work of Babes and Mironescu[242] and Morani,[243] recently reviewed by Krukenberg[244] and Whitman and Stein,[245] is illuminating. These authors showed that a true mesaortitis dessicans, with degeneration and splitting of the elastic laminae of the aortic media may occur. This is the cause and not the effect of the dissection, and may have no connection whatever with the aortic lumen, as in Whitman's remarkable case, in which the wall of the aorta from the base of the heart to 10 cm. above the bifurcation was the seat of a huge mesarterial sac filled with lymph derived from the vasa vasorum, and was formed within the coats of the vessel by the splitting up of their layers without any break in continuity of the intima. Babes and Mironescu further point out that in the dilated aorta such a weakening of the media is likely to be of congenital origin, especially as a bicuspid aortic valve is so commonly associated. Such an explanation would certainly seem to apply to many of the cases of

*New cases in this and Blackford's article.
†Cases in Abbott's article in Osler's System.
‡New cases in this article only.
§New cases in Blackford's article.

rupture in coarctation of the aorta in this series, in which the extent of the tear and the wide dissection of the wall are out of all proportion to the apparent cause. A case in point is Staunig's[153] patient, in whom following upon emotional excitement and in the presence of a very moderate degree of coactation without any collateral circulation, the aortic intima was torn up from a point 3 cm. above the (bicuspid) aortic valve as far as the isthmus. In Wadstein's[144] case, again, the dissection formed nodules beneath the intima of the arch filled with solid blood coagulum, very similar to those described by the above authors in a true "mesaortitis dessicans." Other illustrations might be multiplied from this series.

Localized dilatation of the descending thoracic aorta *immediately below* the stenosis is also common, but this is a direct result of the return of the collateral circulation through the aortic intercostals, for the supply of the lower extremities, and will be considered below in that connection.

Hypertrophy and Dilatation of the Heart.—An increase in the cardiac musculature and cavities, especially of the left ventricle, is extremely common in adult coarctation and was noted in 150 of the 200 cases in the combined series. The presence of cardiac hypertrophy is not, however, an essential feature of even the most extreme degrees of constriction in subjects living to an advanced age. The classic example of this is the oft-quoted case by Reynaud[6] of an old man with a stenosis 1½ lines in diameter and an enormous collateral circulation (see Fig. 11), whose heart was described as perfectly normal, although the brain showed multiple old and recent hemorrhages, apparently the result of an increased pressure in the intracranial circulation.

Other cases reported of a normal heart in this condition are by Otto[4] of a girl, aged seventeen years, with a stricture admitting a pen and no collateral circulation demonstrated, who died with rupture of the aorta; by Dumontpallier,[33] of a woman, aged thirty-nine years, with a stricture 0.012 mm. in diameter and large collateral circulation, dying with failing compensation; by Brunner[97] of a woman, aged thirty years, with complete closure just below the ductus and a large collateral circulation; by Fawcett†[110] and Smith and Targett*[143] (moderate stenosis); by Monckeberg†[120] of a woman, aged twenty-six years, with a stenosis 2 mm. in diameter, dying from rupture of traction aneurysm of descending aorta; by Focken,†[133] in a girl, aged eighteen years, with constriction 3 mm. in circumference, dying from mycotic endarteritis, and the cases by Hansteen§ (3)[199] and Lantinga§[185] in Blackford's series.

*New cases in this and Blackford's article.
†Cases in Abbott's article in Osler's System.
§New cases in Blackford's article.

In view of these facts and the frequent incidence of myocardial and valvular lesions produced by intercurrent infections, it has been claimed by Bonnet,[101] and others,[179] that the fully compensated obstruction in the thoracic aorta makes no extra demands upon the cardiac reserve, and that, therefore, in the absence of such complications no cause for hypertrophy of the heart exists, so that this is never present in uncomplicated coarctation.

The literature does not, however, bear out this contention. Among the 154 cases in which some degree of hypertrophy is stated to have existed, in only 80 were complications or a past history of rheumatism noted. In the remaining 70 cases of hypertrophy there was either no mention of cardiac complications, or it was expressly stated (in no less than 20 of the cases) that these were absent. In these cases no other cause for the hypertrophy of the left ventricle appears to have existed except the hypertension which may reasonably be supposed to have existed in the upper part of the body, although this important diagnostic feature is of course not available from the earlier literature and is too often overlooked in the more recent publications. Among the more convincing instances of hypertrophy of the left ventricle in the absence of any evidence of cardiac complications, or in young and vigorous subjects without history of rheumatism or other damaging infection, may be mentioned such cases as those reported by Erman[54] of a youth of nineteen years with a constriction 5 mm. wide and colossal hypertrophy of the left ventricle, *all valves healthy;* Kriegk's case,[56] a vigorous boy of eleven years, with stricture admitting a bristle, hypertrophy, especially of the left ventricle, and only two aortic cusps, *both healthy and competent.* Follet and Caille's†[129] case, a lad aged seventeen, with blood pressure 210 in the upper and 110 in the lower extremities, dying from cerebral hemorrhage, with a constriction admitting a fine probe, and hypertrophy of the left ventricle, *all valves normal;* Bumke's*[145] patient, a man aged seventeen, with stenosis admitting a sound 1 mm. wide and large collateral circulation, heart greatly hypertrophied, especially on left side, bicuspid aortic valve, *competent not thickened;* Murray's*[149] patient, a man aged forty-eight, with almost complete obliteration of descending arch, great hypertrophy of left ventricle, *all valves healthy;* Fraenckel's Case 2,[156] and the case reported by Meixner,*[175] of a man aged twenty with complete obliteration of the descending arch and good collaterals, dying from rupture of the aorta, in whom the wall of the left ventricle was 3 cm. thick and the aortic valve was bicuspid, *but its two segments were delicate and competent and the other valves were all healthy.*

If the deductions drawn from these descriptions, and other similar ones in the literature be correct, the conclusion is inevitable that while

*New cases in this and Blackford's articles.
†Series in Abbott's monograph, Osler's System.

intercurrent infections play an extremely important and disastrous rôle in coarctation, the coarctation itself, in uncomplicated cases, has also its dangers, by introducing a factor of cardiac strain which may in itself lead to death by failing compensation; so that, even in the presence of a cardiac muscle of perfect integrity, an inadequate collateral circulation with poor vascular tonus in the vessels of its tortuous network does, in the presence of obstruction in the thoracic aorta, itself predispose to cardiac asystole.

The Collateral Circulation.—The gradual development of powerful anastomoses between the arteries above and below the point of constriction in the aorta, which must take place in all patients with extreme stenosis or obliteration of the descending arch surviving beyond infancy, enables life to be carried on with ease and comfort in the absence of intercurrent complications so long as the myocardium remains equal to the task imposed upon it and the heart's valvular mechanism is intact. Fortunately for the early understanding of this subject some of the first cases observed were in dissecting-room subjects in whom the vessels had been injected before the true condition was known, with the result that the finest ramifications of the dilated collaterals were revealed in the later anatomical dissection. This was so in the very first case reported, that by Paris[1] (1791), whose classic description is published in full below, as well as in those by Meckel[5] (1827), and Jordan[7] (1830). In Meckel's case, a peasant, aged twenty-nine years, with a constriction of the aorta just below the ligamentum arteriosum, admitting a very fine straw, who died of rupture of the right auricle of the heart following a strain, the usual large amount of wax was introduced through the innominate artery but disappeared immediately into the dilated collaterals. The injection was thought to be a failure until the abdomen was opened and the astonished observers found the vessels of the lower part of the body to the feet completely filled with the injection mass. From his diagram (Fig. 7-*A*), we can clearly trace the course of the anastomosing vessels. The major part of the new circulation is usually, as in this case, between the superior intercostal arising from the subclavian artery and the first aortic intercostal which springs from the aorta just below the constriction, and the posterior scapular, intrascapular, and subscapular arteries, which, piercing the intercostal spaces from behind, pour the blood coming to them from the ascending aorta by way of the subclavians into the second and sometimes into the fourth aortic intercostals, which form huge, often thin-walled, trunks lying concealed from superficial observation at the back of the thoracic cavities, and which conduct the blood they receive from above from these sources and from the aortic branches of the internal mammary artery into the descending thoracic aorta *below* the constriction (see Fig. 7-*A*). So great is the influx of the returned blood at this point in such cases

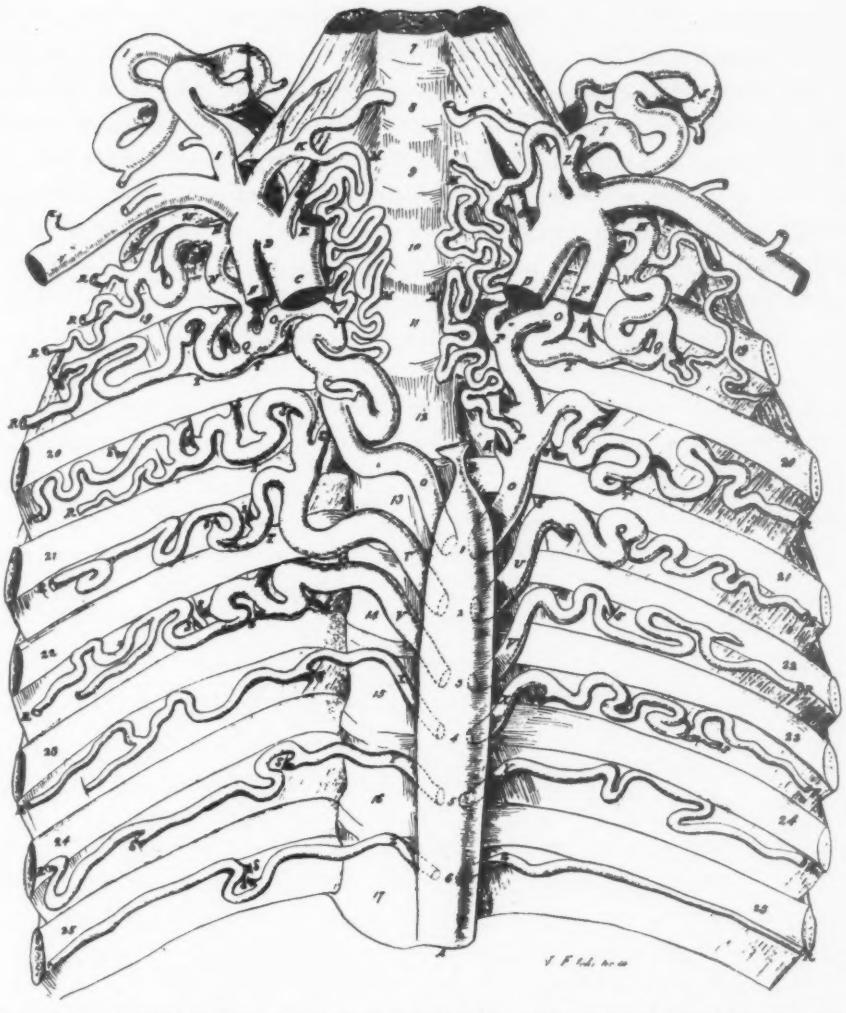


Fig. 7-*A*.—Meckel's case of extreme coarctation with marked development of collateral circulation in a male peasant, aged thirty-five, who died from rupture of right auricle after carrying a heavy load. Drawing showing the anastomoses within the thorax of the vessels above and below the point of stenosis, after injection for dissection. *A*, descending aorta somewhat narrower than usual, especially below, because the amount of injected material was not sufficient to dilate it fully; *B*, narrowing of the aorta to the diameter of a straw; *C*, the innominate trunk; *DD*, subclavian arteries; *E*, right carotid artery; *FF*, internal mammary arteries extraordinarily dilated; *GG*, vertebral arteries; *HHHH*, superior intercostals, as usual branches of the subclavians, but extraordinarily dilated; *IIII*, transverse cervicals and especially their descending branch, which after several large tortuosities which lay under the shoulder muscle and from which some twigs pass into this muscle and the Schulterheber, becomes extraordinarily enlarged and branches along the inner border of the scapula in its course, giving off the numerous large anastomoses (*SS*), with the intercostals; *KK*, inferior thyroids; *LL*, descending cervicals normal on right as branch of inferior thyroid, but on left arising directly from left subclavian; *MMMM*, tortuous anastomes on the right from the inferior thyroid, on left from descending cervical, both bifurcating and opening at *PPPP* into the superior intercostals arising from the aorta; *NN*, the chief anastomoses of all, uniting the first and second intercostals. *OOOO*, upper intercostals from aorta; *PPPP*, opening of branches *NN* RRRR, the cross-section of the intercostal arteries in the region where the lateral branches of the internal mammaries meet them and unite with them as greatly dilated collaterals; *SSSS*, the very numerous anastomoses of perforating branches of the intercostals with the ends of the descending branches of the transverse cervical II; *TTTT*, the excavations in the ribs produced now and then by the arteries; *UU*, second; *VV*, third; *XX*, fourth; *YY*, fifth; *ZZ*, sixth pair of aortic intercostals; *1, 2, 3, 4, 5, 6*, openings of the intercostals in the aorta, not so much dilated as these vessels themselves; *7-9*, the fifth to seventh cervical vertebrae; *10-17*, the eighth upper dorsal vertebrae; *18-25*, the eight upper ribs. (From *Verschliessung der Aorta am vierten Brustwirbel*, By A. Meckel, *Arch. f. Anat. und Physiol.*, given out by J. F. Meckel, 1827, p. 345, Plate V. Figs. 1 and 2.)

that the aorta immediately below the constriction where the three upper aortic intercostals are given off is frequently dilated in a bulbous fashion, as is well shown in Reynaud's[6] case (see Fig. 11), and this dilatation may be so marked as to constitute a true saccular or spindle-shaped diffuse aneurysm (cases of Degen,[47] Eppinger,[50] Monckeberg's Case 1,[119] Reinitz's Case 2[147] with atheromatous walls projecting to the left and which in one instance ruptured into the left bronchus (Leudet[35]), and in two others into the left pleura (Fawcett, Obs. 5[109]; Monckeberg[120]). Even where this localized increase in diameter is not marked, the aorta just below the constriction at the entrance of the collateral blood often shows a patchy atheroma, which may act as a predisposing factor for the formation of a dissecting aneurysm with spontaneous rupture in this situation or

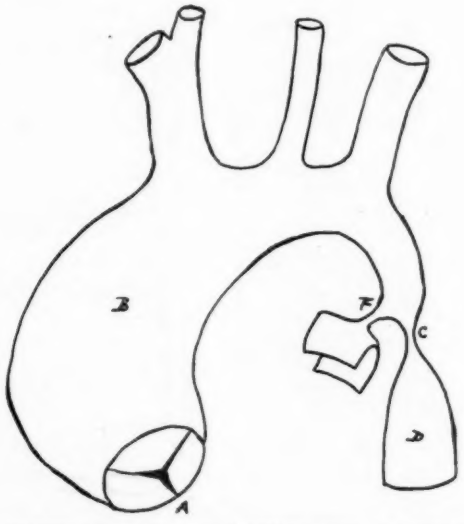


Fig. 7-*B*.—Outline drawing of aorta in Meckel's case of extreme coarctation showing: *A*, aortic valve; *B*, dilated ascending aorta; *C*, stenosis of descending arch immediately below; *F*, the obliterated ductus; *D*, the descending aorta, moderately dilated immediately below the constriction. (From *Verschliessung der Aorta am vierten Brustwirbel*, By A. Meckel, *Arch. f. Anat. und Physiol.*, given out by J. F. Meckel, 1827, p. 345, Plate V, Figs. 1 and 2.)

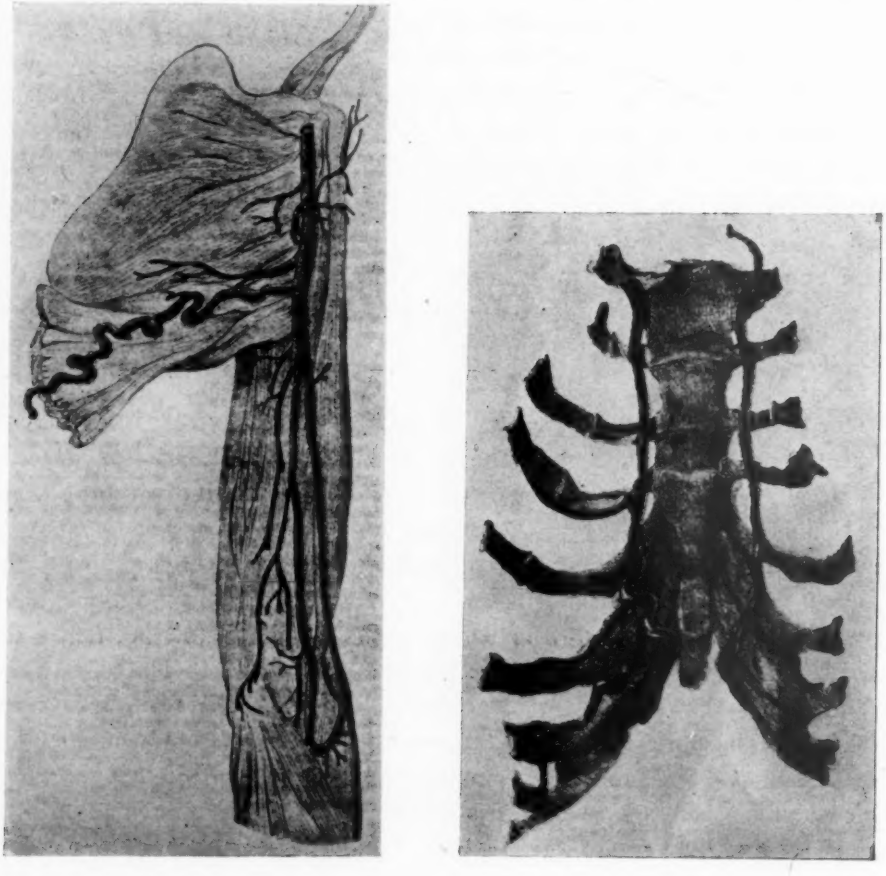
for the development of a mycotic aneurysm as in Focken's two cases[132, 133] and those by Fawcett,[115] Smith and Targett,[143] Smith and Hausmann,[135] Reifenstein,[134] Beneke,[176] Libman,[180] and Mackenzie.[181] The aorta resumes its normal, or a diminished, caliber at the level of the fourth or fifth intercostal arteries which usually take no part in the anastomosis, but supply their own district from the blood which reaches them through the aorta by this circuitous route. The fact that the great volume of blood in patients with this intercostal type of circulation is returned directly into the thoracic aorta from collaterals which course chiefly within the thorax or are largely concealed beneath the heavy muscles of the back, explains what appears at first sight to be a bizarre and curious phenomenon, in that the most ex-

treme degree of constriction with a perfectly developed collateral circulation may exist, without any of those manifestations yielded by tortuous pulsating vessels beneath the skin of the upper part of the body or diminished femoral pulse below, which constitute, when present, such important diagnostic features. It also explains why, in such cases, when nothing may be made out anteriorly by inspection or palpation, careful examination of the *back* of the thorax will often detect signs of a pulsating vessel between the shoulders (as figured by John T. King[208]), or an arterial murmur at, or a little above, the lower angle of the scapula produced in the dorsalis scapulae or the subscapular branch of the brachial artery. The place of origin of this murmur in the latter situation is beautifully shown in the picture of this injected vessel from Jordan's[7] case (1830) (see Fig. 8-*A*) of a young man, aged twenty-one years, with obliteration at the ductus, who died from rupture of the ascending aorta. In other cases the clue to the diagnosis was obtained from the presence of a rough murmur along both sternal borders produced in the dilated internal mammaries (Parkes Weber[221]).

The latter arteries frequently supply another source for the collateral circulation from the subclavian arteries to the lower extremities which in some instances appear to be the predominating one, these vessels becoming continuous with the epigastric branches of the external iliacs over the abdominal wall. When this path is well developed, the signs may be extremely striking, a loud holo- or postsystolic murmur being audible along either parasternal line, while the epigastric arteries may stand out like whipcords (as in Kolisko's Case 3[160] and those by Tiedeman,[19] Focken,[132] Sella[124] and Hart[167]); or from their tortuous course they may resemble a cirsoid aneurysm beneath the skin of the abdomen (cases by Libman,[99] Decker,[91] and Reinitz[146]). This second route is, however, less constant, and as Bonnet has pointed out, it is much less important for the adequate supply of the lower extremities than is the deep supply by the more direct route into the thoracic aorta by means of the aortic intercostals; so that the presence of distinctive signs over the internal mammary and deep epigastric arteries is again not an essential part of the diagnostic picture. This was pointed out by Graham[2] in 1814 in his account of the second case on record, a boy of fourteen years with complete obliteration, in whom the dilatation of the aorta just beyond the constriction and the absence of any change in the epigastric arteries showed that the circulation had reached the femoral artery from the thoracic aorta and not by way of the abdominal wall.

Both these paths of collateral circulation were prominently established in a number of the most typical cases, as in those figured here from Reynaud's and Jordan's articles, and in Parkes Weber's[183] and

our own patient.[136] It is of interest also to remember that the circulation for the lower part of the body is in either case derived practically entirely from the subclavian arteries, which assume a great size and pulsate widely, while the carotids are usually relatively little changed. Nevertheless the peripheral branches of the latter suffer under the effect of the high intracranial blood pressure that com-



A. *B.*

Fig. 8.—Drawings from Mr. Jordan's case showing details of the collateral circulation in areas of much clinical significance.

A, The dilated and tortuous subscapular branch of the brachial artery passing beneath the scapula to pierce the intercostal space posteriorly on its way to join the upper aortic intercostals. *B*, The dilated internal mammary arteries passing down behind the sternum to meet the deep epigastric arteries over the abdominal wall and sending branches through the anterior intercostal spaces to the upper aortic intercostals.

Male, aged twenty-one years with atresia of descending arch, died of rupture of the aorta. (From A Case of Obliteration of the Aorta with Disease of the Valves, By J. Jordan, *North of England Med. and Surg. Jour.*, 1830, i, 101.)

monly prevails, so that the cerebral arterioles are frequently atheromatous and are sometimes the seat of multiple aneurysmal dilatations, in which the new conditions of the circulation may have some causal connection.

The vertebral arteries, being the first given off from the subclavians,

are liable to share in the elongation and dilatation of the collateral vessels. The only instance within our knowledge in which this was of clinical significance is Haberer's[148] case, in which a kinked and tortuous branch of the anterior spinal artery produced a compression myelitis of the spinal cord at the level of the second dorsal vertebra, followed by the development of a paraplegia three months before death.

Another result of the altered circulation that is of great importance for the diagnosis of these cases during life, and that requires some elucidation here is the *retardation* and *diminution* or even *absence* to the palpating finger *of the femoral pulse.* This sign, in combination with evidence wheresoever obtained, of dilated pulsating vessels or arterial murmurs in the upper part of the body is pathognomonic of coarctation. It is, however, *not constant,* either in its presence or in the extent of the diminution, which seems to bear no relation to the degree of the stenosis, for this may amount to actual obliteration without noticeable change in the pulsation in the femorals, while, on the other hand the femoral pulse may be almost imperceptible in the presence of a relatively moderate degree of coarctation. This variability, and the fact that it is difficult to understand how the actual volume of blood could be seriously lessened while the nutrition of the lower extremities is fully maintained (as is the case in most of these subjects), is at first sight very perplexing. We owe our understanding of this apparently contradictory phenomenon to Bonnet's brilliant exposition of the physiological facts as they are known to exist. He pointed out that the sensation imparted to the palpating finger from the pulsating vessel is not dependent on the volume of blood in the latter but on the more or less sharp repercussion of the wave that passes along it as the blood is propelled into the aorta. When this is sudden and abrupt, as under normal conditions, the tactile sensation imparted is that of a forcible pulsation; when, however, the blood reaches the aorta through a large series of small collaterals, the ascent will be more gradual and the pulse below it correspondingly weaker. In other words the *sphymographic tracing* supplies the key to the facts, by showing that it is not the *volume* of blood in the femoral artery that is reduced or altered by the coarctation, but the *pulse wave* that rides upon it to meet the palpating finger, which shows a more gradual line of ascent and a more rounded upper curve distinctly retarded behind the abrupt rise of the normal radial tracing. The more devious the paths by which the blood returns to the aorta, the more gradual will be the curve in the femoral tracing and the weaker the femoral pulse will feel; while where the blood is returned immediately below the constriction through the hugely dilated upper intercostals (see

Fig. 7-*A*), the curve may be so abrupt as to resemble closely the normal, and the femoral pulse will be almost as strong to palpation as that in the upper extremities. It follows, therefore, that the force of the pulse under palpation depends, not upon the degree of constriction of the aorta, but on the directness of the path taken by the blood back into the thoracic aorta, and that the most extreme cases of coarctation may be the most difficult to diagnose, simply because the blood has taken, in accordance with known physiological laws, the shortest possible path back into the stream entering the abdominal aorta. These

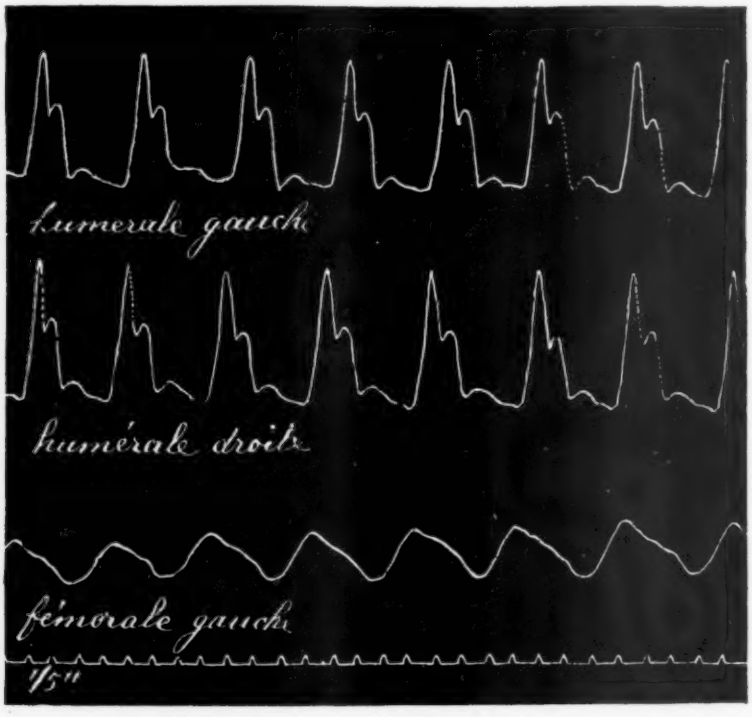


Fig. 9.—Sphygmographic tracings from the left brachial, right brachial, and femoral arteries in a patient diagonosed clinically as coarctation of the aorta, in whom, the left radial pulse was stronger than the right, and the difference in the systolic pressures in the upper and lower extremities was 70 mm. (Note the difference in form and amplitude of the brachial and femoral tracings; and the retardation of the latter. (From Laubry and Pezzi: *Traité des Maladies Congenitales du Coeur*, J. B. Ballière et fils, 1921, p. 263, Fig. 94.)

facts are clearly presented also by Scheele[212] and Laubry and Pezzi[207] with tracings showing the characteristic slow ascent and retardation of the femoral pulse as compared with the normal radial (see Fig. 9).

A marked *difference between the systolic blood pressure* in the upper and lower extremities (the latter being as abnormally low as the former is unusually high, while the diastolic pressure remains but little changed), is probably the most important single sign of the presence of the obstruction in the thoracic aorta and the new conditions of the circulation. For reasons not altogether clear at the present time, this

sign is less subject to variations than is the change in the femoral pulse. Routine observations are lacking, but we believe this difference is almost always present in well-compensated cases of extreme coarctation. The two pressures should, therefore, always be taken in suspicious cases, and conversely, in cases of hypertension, the femoral pressure should always be compared with a view to excluding this possibility.

(*To be continued.*)

TWO TO ONE RIGHT BUNDLE-BRANCH BLOCK

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AND

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TWO to one right bundle-branch block is very rare. We have found no instance of it in the literature, and no such record has occurred among 13,875 electrocardiograms of 8,545 subjects at the Massachusetts General Hospital in thirteen years (1914 to 1927). It consists of the alternation in normal sino-auricular rhythm of normal and abnormal ventricular complexes, the abnormal QRS waves representing fully defective conduction in the right bundle-branch. It is comparable to two to one auriculoventricular heart-block, when the entire bundle of His, and not alone its right branch, fails to conduct the auricular impulse to the ventricles at every other beat.

In a personal communication to us, Dr. Nils Stenström,[1] of Stockholm, Sweden, has written of one instance of two to one right bundle-branch block that has come to his notice. This case of his he has already reported for other reasons, but the electrocardiograms showing this particular anomaly, he has not yet published. He has given us permission to record the curve in question (Figs. 1 and 2). Unlike our case, however, this patient of Dr. Stenström's showed the two to one bundle-branch block only as a brief transitional event, occurring between relatively normal bundle-branch conduction and complete right bundle-branch block. When the heart rate was speeded up by exercise, conduction through the right bundle-branch became defective in every beat. When it was slowed by vagal pressure, intraventricular conduction became practically normal. The two to one right bundle-branch block occurred at times for a few beats in the period intervening between vagal pressure and exercise.

Intraventricular block is common, especially lesser degrees and complete right bundle-branch block. Not infrequently electrocardiograms have been published, illustrating variations in degree of the block or even alternating right and left bundle-branch block,* but, as noted above, two to one block in either bundle-branch is very rare. An excellent example of two to one right bundle-branch block, later becoming complete, has been encountered by us, and this case we are herewith reporting.

CASE REPORT

C. F. M., white, a hardware merchant, aged sixty-five years.

Diagnosis.—(1) Hypertensive cardiovascular disease, (2) angina pectoris, and (3) right bundle-branch block.

*See references [2], [3], [4], [5], [6], [7], [8], [9], [10], [11], [12], [13], [14], [15].

Complaint.—Paroxysmal pain in chest and arms.

Family History is not significant.

Previous Medical History.—Patient has always been healthy until recent years. He had typhoid fever at the age of forty, and lost his left hand in a railroad wreck thirty-six years ago. For many years he has been a sufferer from chronic constipation and indigestion. He denies having had venereal disease. Three years ago he had a quite severe attack of bladder trouble, with difficulty in voiding for several days. Some months later his systolic blood pressure was found to be 210 mm. of mercury during a life insurance examination.

History of Present Illness (December, 1926).—The present symptoms appeared in September, 1926, while milking a cow. At that time he had a brief, severe substernal pain, radiating to shoulders and arms and accompanied by oppression in chest. Some days later another attack occurred while he was waiting on a customer in his store. From that time up to the present he has had numerous attacks of this paroxysmal pain. All attacks have been short, none exceeding five minutes in duration. The pain is diffuse but maximal in the precordium and substernal region and radiates to both shoulders and arms, more intensely to the right. There is marked oppression accompanying these paroxysms. The pain is definitely brought on by exertion and also by emotional excitement. He is uncertain as to its relation to exposure to cold and to overeating. In general the pain occurs more often at night. He has found it necessary to walk very slowly; climbing steps has become impossible if he wishes to avoid bringing on paroxysms of pain. The attacks are relieved very definitely by amyl nitrite. He is somewhat more short of breath and weaker than he has ever been before. No symptoms or history could be elicited by close questioning of the patient of any attack which might be interpreted as coronary occlusion.

Physical Examination (December 1, 1926) shows a well-developed and nourished man with ruddy countenance, and normal mentally. The eyes are negative in every respect but for, perhaps, a slight degree of retinal arteriosclerosis. The tongue is clean. There is a moderate degree of pyorrhea around both upper and lower teeth. The thyroid gland is not enlarged. There is no abnormal pulsation, either arterial or venous, in the neck. The lungs are negative. There are no râles at the bases.

Heart.—Percussion reveals a moderate degree of cardiac enlargement. There is a dominant rhythm with no premature beats; no murmurs. The aortic and pulmonic second sounds are about equal. There is quite a striking reduplication of the second sound with every alternate systole, not found with any of the intervening beats. This finding was repeatedly noted. Blood pressure in the left arm, reclining, 200 to 206 mm. of mercury, systolic; 100 to 104 mm., diastolic. Above a cuff pressure of 110, all sounds heard at the elbow are sharp and equal. Below a cuff pressure of 110, there is an alternation of sharp beats and dull ones. This alternation in quality of the sounds heard at the elbow is constant.

The abdomen is slightly distended with gas; otherwise negative. Neither liver nor spleen is palpable. The skin, extremities, nervous system (reflexes) and back are all normal except that a pedunculated lipoma about the size of a cherry is observed over the lower right thorax.

Laboratory Data.—Urine, December 1, 1926, shows a neutral reaction, a specific gravity of 1.015, no albumin or sugar, and negative sediment. The red blood count is 3,440,000; hemoglobin 80 per cent, and white blood count 6,200. Blood urea nitrogen is 15 mg., and creatinine 1.66 mg. per 100 c.c. of blood. The phenosulphonephthalein output is 34 per cent the first hour and 13 per cent the second hour.

Electrocardiogram December 1, 1926 (Fig. 3) shows normal rhythm, rate 105, with every alternate impulse blocked in its passage along the right bundle-branch.

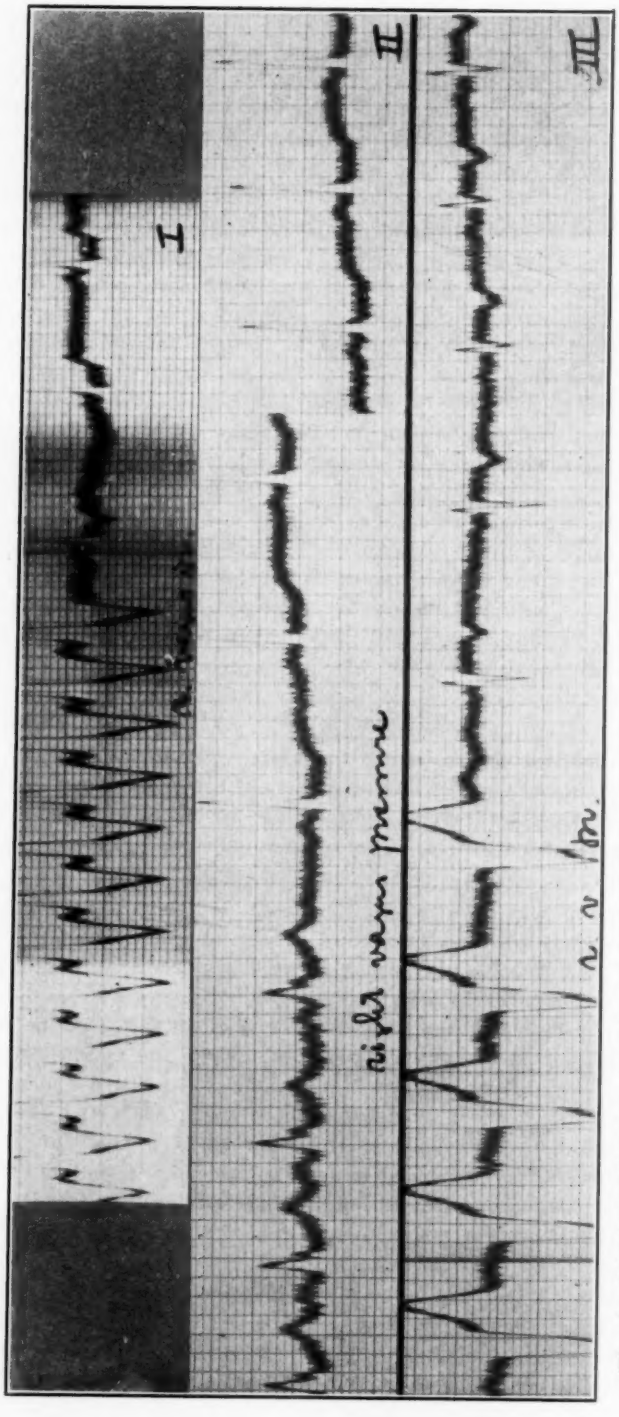

Fig. 1.—Electrocardiogram (Leads I, II, and III) of Dr. Nils Stenström's patient, showing at onset tachycardia and right bundle-branch block, resulting from exercise, and, following this, slowing of the heart rate and normal intraventricular condition occurring with right vagal pressure. Time: 0.1 second; voltage: 1 mm. $=10^{-4}$ volt. (x2/3.)

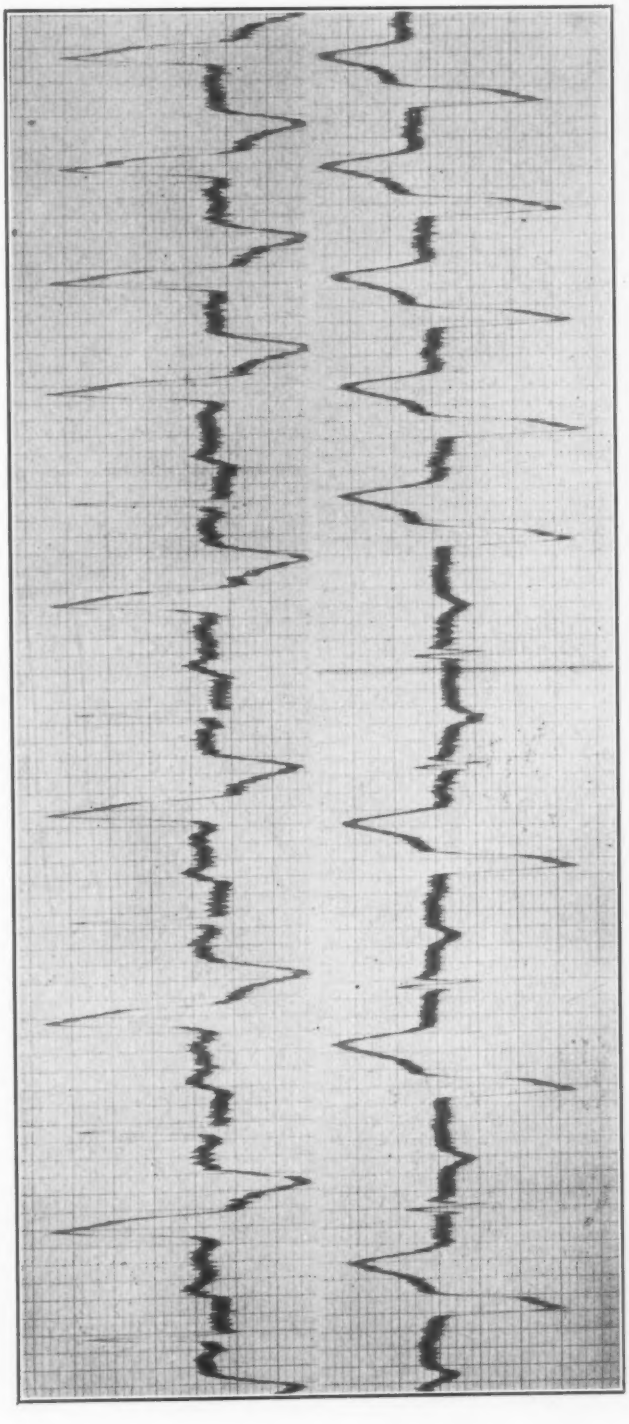

Fig. 2.—Electrocardiogram (Leads I and III) of same patient, showing at first a transient two to one right bundle-branch block, which later became complete. Time: 0.1 second; voltage: 1 mm.=10^{-4} volt. (x2/3)

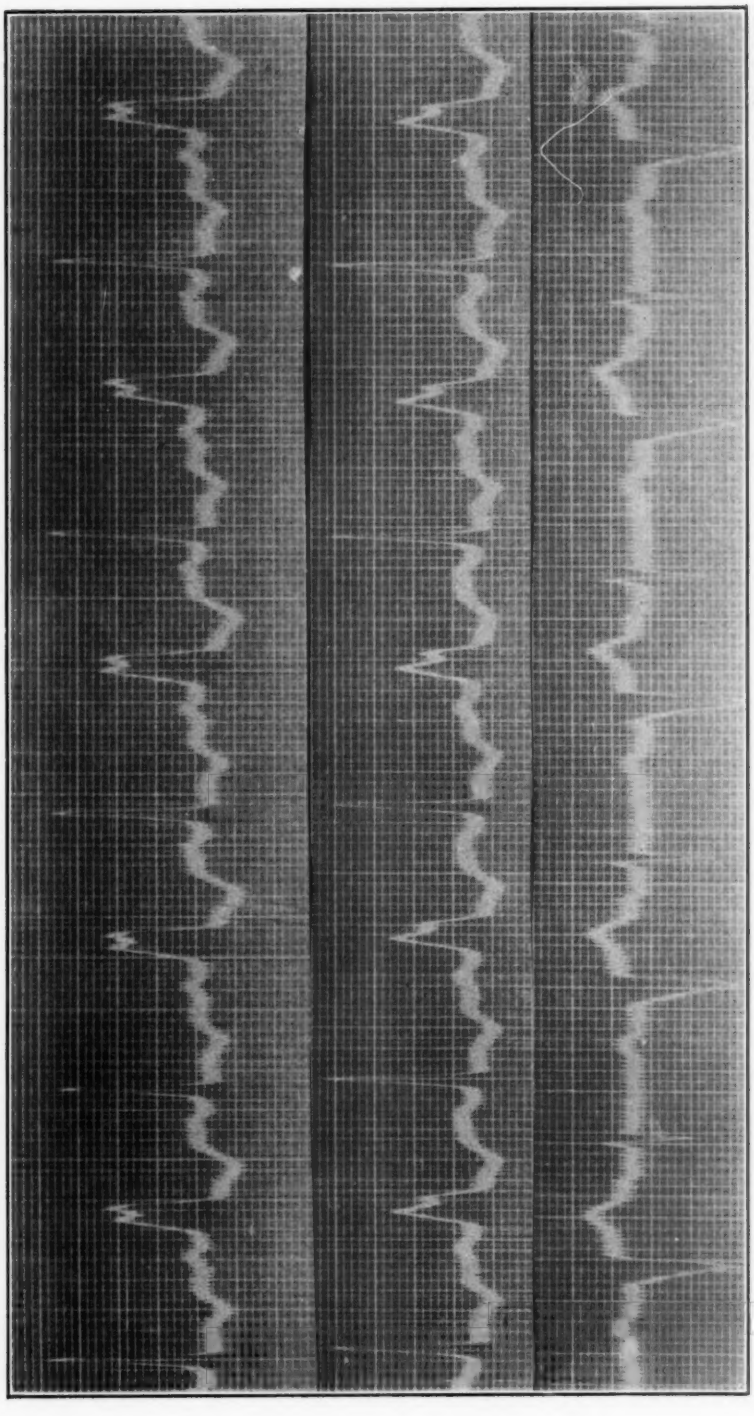

Fig. 3.—Electrocardiogram (Leads I, II, and III) of case here reported, showing (Dec. 1, 1926) two to one right bundle-branch block. With normal sino-auricular rhythm, alternate ventricular complexes are normal and abnormal. Note also the inverted T-waves following the normal QRS waves in Leads I and II. Heart rate 105. Time 0.04 second; voltage: 1 mm.=10^{-4} volt. (x2/3)

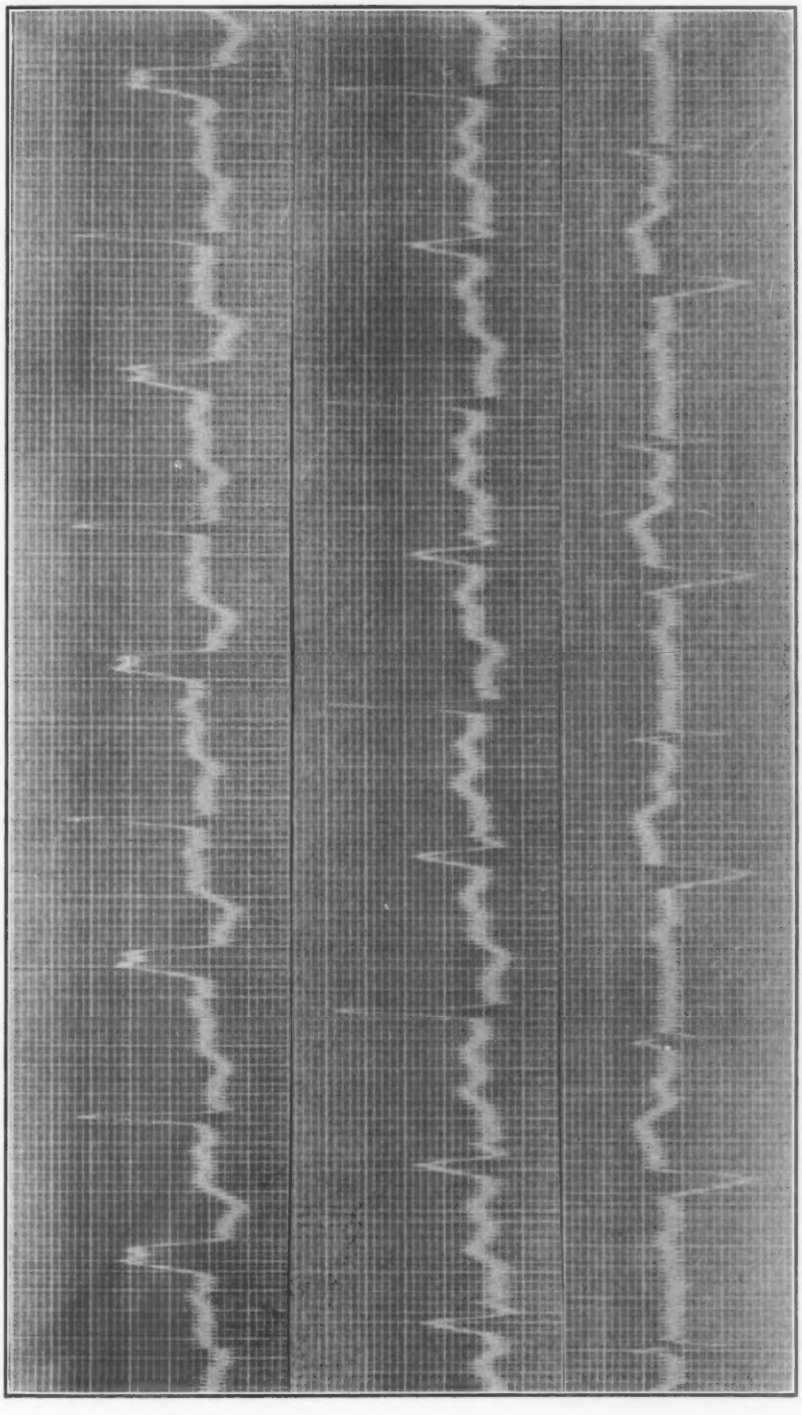

Fig. 4.—Electrocardiogram (Leads I, II, and III) of same case three days later, showing the same two to one right bundle-branch block, except for two contiguous blocked impulses at the beginning of Lead II. Time: 0.04 second; voltage: 1 mm.=10^{-4} volt. (x2/3)

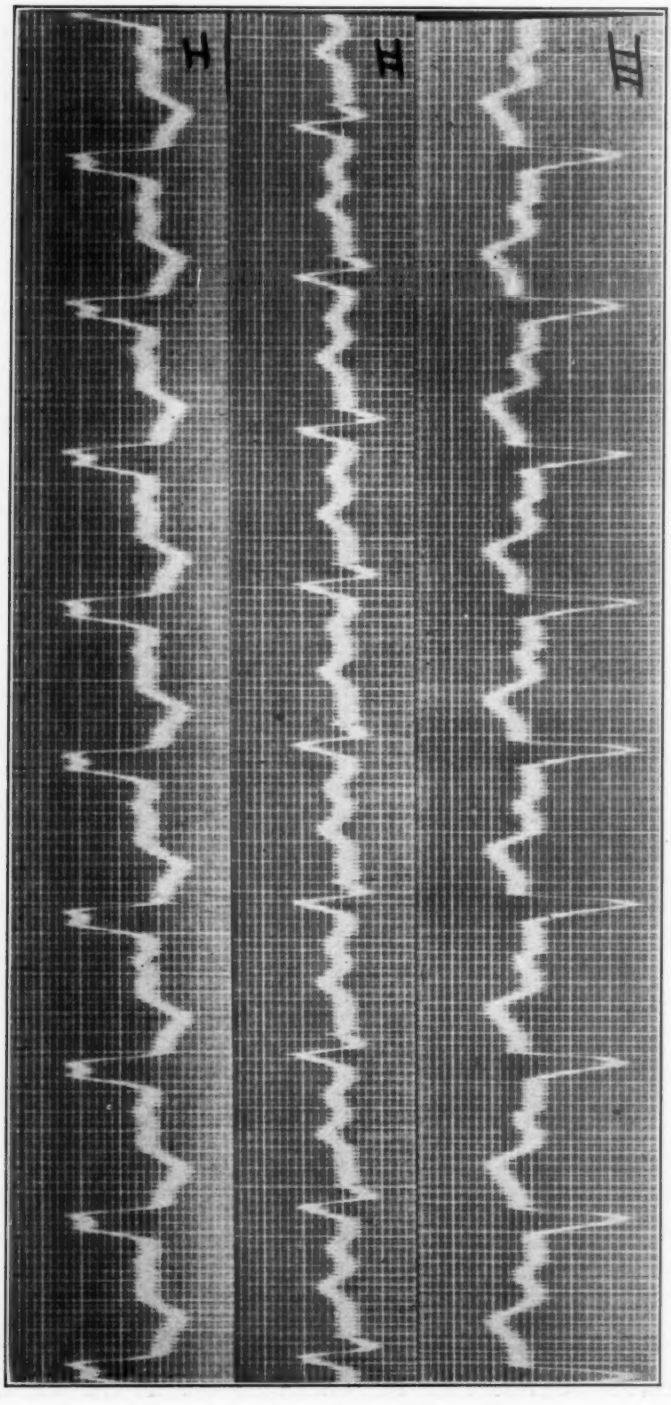

Fig. 5.—Electrocardiogram (Leads I, II, and III) of same case two days later (Dec. 6, 1926), showing complete right bundle-branch block constantly. Rate 95. Time: 0.04 second; voltage: 1 mm. = 10^{-4} volt. (x2/3)

Intermediate impulses pass normally over both branches. There is slight left axis deviation. The T-waves are inverted in Leads I and II. The tracing of December 4 (Fig. 4) shows identically the same conditions (rate 95), save only that two contiguous blocked impulses are noted. The tracing of December 6 (Fig. 5) shows normal rhythm, rate 95, and complete right bundle-branch block with no normal QRS complexes remaining.

SUMMARY

A case is reported showing by electrocardiogram at first two to one and later complete right bundle-branch heart-block. No other similar case has been found recorded in the literature, but curves of another patient showing the condition and obtained by Dr. Nils Stenström, of Sweden, are also published herewith.

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THE DIAGNOSIS OF CHRONIC MYOCARDITIS WITHOUT CARDIAC INSUFFICIENCY

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THE condition recognized clinically as "chronic myocarditis" includes a variety of pathological lesions which can be differentiated at the post-mortem examination. Under this term the clinician accepts various degenerative and inflammatory processes which damage the heart muscle, causing a change in the muscle cell, and which are usually associated with an increase in connective tissue. These lesions may be the result of such diverse factors as metallic poisoning, arterial changes, infections, such as diphtheria, gonorrhea, syphilis, rheumatism, etc.

Attention is usually first directed to the possibility of this diagnosis by a careful history which indicates some degree of *cardiac insufficiency* elicited by the physical or emotional stress of daily life. At first such symptoms may appear only as an occasional manifestation following some exceptional physical exertion, such as running for a car or carrying a heavy bag, or they may follow an unusual emotion, such as is aroused by the death of a friend or sudden financial reverses; they often make their presence evident after loading the stomach with a large or injudiciously selected meal. Later the evidences of a heart failing to maintain an adequate circulation are more conspicuous and continuous and are evoked by the more common activities, such as stair-climbing, household duties, and the turmoil and worries of business and family life. Almost invariably the diagnosis of "chronic myocarditis" is first suggested by signs or symptoms of cardiac insufficiency.

There is, however, a group of cases in which, notwithstanding the absence of any evidence of cardiac insufficiency, the diagnosis of myocarditis may be made with a considerable degree of assurance.

There are a number of facts which make it probable that such cases occur and can be detected. *First,* it is obvious that myocardial changes frequently begin as small limited lesions; these are often found at the post-mortem examinations of persons dying from causes other than circulatory failure. These lesions are so small or are so situated in the cardiac tissues that during life they have failed to compromise the work of the heart. On the other hand, while they may not have been large enough to interfere with the ability of the heart to perform its work efficiently, they may have been so placed as to cause abnormal physical signs or to furnish evidence, elicited with instruments of precision, which marked their presence indubitably. *Second,* there are certain symptoms which are so frequently

associated with pathological changes in the heart muscle, that in their presence one is at once warranted in assuming that there is a defect in the tissues of the heart even in the absence of evidence of functional incapacity. Extrasystoles which are frequent and persistent and which arise from several foci in the heart muscle; continuous auricular fibrillation; defective conduction, and alternation of the pulse are with very few exceptions due to myocardial damage. Some of these may be found by the ordinary methods of physical examination; at times they can be detected only by employing graphic methods. *Third,* when one has the opportunity of following patients belonging to this group over a long period of time, he will not infrequently find his diagnosis verified by the development of other symptoms including those of outspoken myocardial insufficiency, and a post-mortem examination may ultimately afford complete proof of earlier diagnosis of disease of the heart muscle.

I can best illustrate the type of individuals who belong to this category by giving a brief outline of several of these cases.

CASE 1.—An electrical engineer, twenty-nine years of age, came to secure my influence in obtaining a commission in the army (1917) which had been refused on account of an irregular heart action. Except for measles and chickenpox as a child and numerous attacks of tonsilitis, he had never been ill. His habits were good. He could not believe that he had any serious heart affection since, with the exception of a consciousness of his irregularity, he felt in perfect health. He had always indulged in hard and prolonged exercise and just before his first visit to my office had played five sets of singles in tennis without shortness of breath or weariness. The physical examination revealed nothing abnormal except a complete irregularity of the heart with a rate of 76. Various observations to determine the capacity of the heart for work, including exercise response and vital capacity determinations, showed no evidence of cardiac insufficiency. The heart was normal in size, verified by fluoroscopic examination; there was complete irregularity but no murmurs. An electrocardiogram showed the presence of auricular fibrillation, and a diagnosis of myocarditis was made. I have seen this patient recently (1927); his heart is now slightly enlarged, and the irregularity persists. There has been no material increase in the heart rate. He has cut out his most strenuous forms of exercise, but has no shortness of breath with ordinary exertion; he admits getting a "bit tired" when he climbs a hill. In spite of this remarkable record of cardiac efficiency, I feel that the diagnosis of chronic myocarditis is justified, and I believe it is only a matter of time when the symptoms of a failing heart will become evident.

CASE 2.—A man, sixty-seven years of age, first consulted me in 1916, because he was conscious of an irregular heart action and was curious to know its cause. A most careful history failed to elicit any story of infection of any kind. He had for many years held an important professorship at one of our great universities, and in the summer months had devoted himself to archaeological exploration in the Rocky Mountains, work requiring a great expenditure of physical energy at considerable altitudes. He had been conscious of the cardiac arrhythmia for some months, but neither his history nor physical examination revealed any symptoms of cardiac insufficiency. Examination showed a heart that was completely irregular, a rate of 80, without enlargement and without murmurs. The electrocardiogram

confirmed the presence of auricular fibrillation. A diagnosis of chronic myocarditis was made. His activities were curtailed. Later as his heart became more rapid, he was given small doses of digitalis. He showed no other signs of cardiac insufficiency until two years later when he developed slight shortness of breath on exertion. From this time on in spite of the most careful regulation of his mode of life and efficient digitalization, the signs of a failing heart became more and more apparent, with marked dyspnea, edema of the legs, enlarged liver, etc. He died in 1924. The autopsy showed a general arteriosclerosis, with pronounced involvement of the vessels supplying the heart muscle. The ventricular wall showed a marked myocarditis with areas of extreme atrophy and other areas of newly developed muscle tissue.

CASE 3.—A woman, sixty years of age, came to me for advice in 1919. Her chief complaint was epigastric pain without radiation. As a girl she had suffered from a severe and prolonged attack of rheumatic fever. She had always been troubled with an obstinate and extensive psoriasis, and in recent years a great deal of gastrointestinal disturbance. Her pain was thought to be due to gallstones, and an operation had been advised. Unless the pain be regarded as such, neither the history nor physical examination afforded any evidence of cardiac insufficiency. Her heart was of full size but not definitely enlarged; the sounds were of good quality; there was a faint systolic murmur heard in the fourth space just to the left of the sternum, and an extrasystole was occasionally detected. The electrocardiogram showed a curve presenting all the classical features of a block of the right branch of the bundle of His. It was quite clear that she had a serious myocardial defect, and she was advised accordingly. The diagnosis was verified by the subsequent development of symptoms of cardiac insufficiency, and she died of heart failure in 1924.

The above histories will serve to indicate the type of case in which the diagnosis of "myocarditis" is justified even in the absence of evidences of cardiac insufficiency. In the second and third cases the earlier diagnoses have been verified by subsequent events, and in one an autopsy made the proof complete.

In addition to the abnormal features which have been enumerated, the probability of a correct diagnosis may be made more secure by a history of a definite etiological factor or by the presence of other symptoms and physical signs, such as precordial pain, enlargement of the heart, a murmur, bradycardia, tachycardia, unstable heart rate, or a change in the heart sounds, such as reduplication and gallop rhythm. On no one of these alone would one be justified in hazarding a diagnosis of heart muscle defect, but they may serve as corroborative evidence of such a condition.

It should be emphasized that this group of cases is a small one. Evidence of an inadequate circulation is, as a rule, an early feature in those having continuous auricular fibrillation, conduction defects, and the other abnormalities mentioned above. When the myocardium is damaged, the development of cardiac insufficiency is usual. In a limited number of cases, however, it is possible to make a diagnosis of a defect of the muscle before the heart begins to fail in the efficient performance of its work.

BLOOD VESSELS IN THE VALVES OF NORMAL HUMAN HEARTS*

From a Study of 700 Human Hearts

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IT IS obvious that unless the conception of the existence of blood vessels in the valves of some human hearts can be established beyond a reasonable doubt, they cannot be seriously considered as a contributing factor to the pathogenesis of valvular endocarditis. With this in mind two of us reported the results of our investigations on 85 human hearts[1] in which very strong evidence was brought to bear to show that blood vessels of noninflammatory origin do at times exist in the valves of human hearts after birth.

For purposes of review and because it is pertinent to our present discussion we present the following summary of the main points which led us to this belief:

1. The constant and characteristic architecture of the blood vessels found in the valves as opposed to the bizarre inconstant appearance of granulation tissue blood vessels.

2. The origin of these blood vessels from the anastomotic arch which springs from the main coronary arteries, and not from the neighboring capillaries as would be expected in granulation tissue.

3. The characteristic venous return from the valves into the coronary sinus.

4. The histological demonstration of arterial and venous structure of these vessels as shown by the presence in the former of an intima, internal elastica, and a muscular media, and in the latter of a more delicate muscular wall.

For reasons which we outlined in the report mentioned, this summary deals specifically with the aortic cusp of the mitral valve. It is well to point out at this juncture that a number of these valves were the seat of an endocarditis, but in spite of this fact our investigations led us to the conclusion that the fully formed blood vessels found in these valves did not owe their origin to inflammatory granulation tissue but had existed prior to the inflammation. The results of a further study of 615 additional human hearts have been thus far in full agreement with this conclusion.

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In this paper we propose to describe the blood vessels as they occurred in the valves of 14 normal human hearts which are of unique interest both in themselves and in the support which they lend to our view. Before doing so, however, it is important to point out that we have been able to subdivide architecturally the blood vessels (arteriae valvulares) as they occur in the aortic cusp of the mitral valve of the human heart into two types; namely, (1) complete form and (2) incomplete form:

1. The complete form (Fig. 1) (representing probably a persistence of the fetal type of architecture) is made up of an anterior, a posterior, and an intermediate branch which, after coursing for some dis-

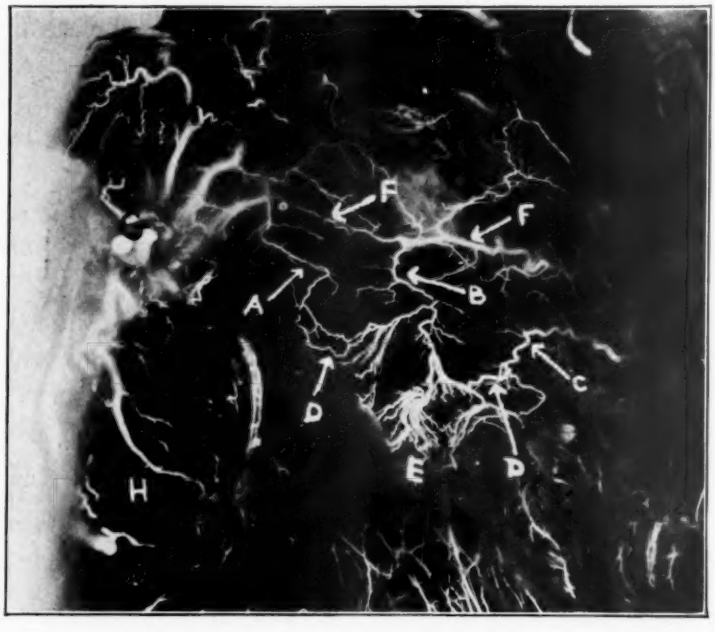

Fig. 1.—Photograph of injected and cleared human heart, showing the vasculature of the aortic cusp of the mitral valve in the "complete" type.

A, anterior descending branch; *B*, intermediate descending branch; *C*, posterior descending branch; *D*, vascular arch (double); *E*, brushwork of vessels to line of closure; *F*, arteria anastomotica auricularis magna; *G*, left coronary artery; *H*, anterior wall of left ventricle.

tance toward the closing edge of the valve, are united by an arcuate vessel. The latter frequently gives rise to a brushwork of finer vessels which descend toward the line of closure. The anterior, posterior, and intermediate branches almost invariably arise from an anastomotic loop (arteria anastomotica auricularis magna) which runs in the interauricular septal musculature to join the left and right coronary arteries or their branches.

In a separate publication Kugel[2] has shown that this arteria anastomotica auricularis magna is a vessel which is found with great constancy in human hearts. Branches descend from this arch toward the

base of the aortic cusp of the mitral valve. In nonvascularized valves these vessels (evidently aborted arteriae valvulares) terminate in the auricular musculature near the base of the valve and often form an arch at this site. At times these vessels, with or without the arch, enter the substance of the valve and terminate either near the base, or at any site between this and the closing edge of the valve. Thus, it would seem that the arteriae valvulares of the vascularized valves either represent prolongations of the constantly occurring aborted vessels coming from the arteria anastomotica auricularis magna or are a persistence of the vessels as they occur in the embryonic valves, i.e., without having undergone complete regression.

2. The incomplete forms present a series of gradations in regression (or development). It is noteworthy that in this incomplete form the arch is invariably absent. Indeed, it is largely on this basis that we have made our division into the two types.

In the 85 hearts previously reported 7 showed the complete form of mitral vasculature. All 7 hearts showed a coexisting rheumatic valvulitis. As stated before, despite the coexistence of valvulitis we were unable to depart from the conclusion that the larger vessels were not of an inflammatory origin. The interest and importance of the 14 hearts which we will now describe lies in the fact that 6 of these presented the complete type of mitral valve vasculature, and 8 presented the incomplete type, but that in no respect either clinically or pathologically could evidence be found that the valves had been subject to an inflammatory lesion or that the myocardium had at any time been the seat of any inflammatory pathological process.

In our pathological examination of the hearts, numerous sections were taken from auricles, ventricles, valves, and base of the aorta, particularly at those sites which are the most frequent seat of Aschoff bodies. The hearts were fixed in alcohol whenever rheumatic disease was suspected. The sections were stained with hematoxylin and eosin, with the Unna-Pappenheim stain, with van Gieson's stain, with the MacCallum-Goodpasture stain, and with Weigert's elastic stain. We particularly searched for Aschoff bodies, Bracht-Waechter lesions, MacCallum's auricular lesions,[3] the lesions described by Klotz,[4] by von Glahn and Pappenheimer,[5] and for any specific or nonspecific foci of inflammation. In the general examination of the hearts we also looked for pericarditis and for the commissure lesions described by Lewis and Grant.[6] A complete examination was made of all the parenchymatous organs with emphasis on the kidneys and spleen.

To make clear some points of discussion which follow, one must state that out of 700 hearts obtained at autopsy the 14 described in this report were the only normal hearts, as judged by the above standards, showing blood vessels in the valves.

As is seen from the foregoing, the hearts which we eliminated from this series fall into two groups:

A. Those showing clinical or pathological evidence of endocarditis, past or present.

B. Those in which were found small myocardial focal accumulations of leucocytes, small lymphocytic foci in the valves (Fig. 2), etc. In none of these hearts was there found pathological evidence of such a nature as to make it probable that the blood vessels seen in these valves were secondary to inflammation. Moreover, with one exception, so far as we were able to determine, there was no previous history of rheumatic fever, chorea, arthritis, scarlet fever, or repeated attacks of tonsillitis. This, therefore, represents an *intermediate group* which perhaps does not justify its complete elimination from statistics intended to show the relative number of hearts possessing arteriae valvulares in which the occurrence of these vessels can be reasonably attributed to an inflammatory lesion in the valve.

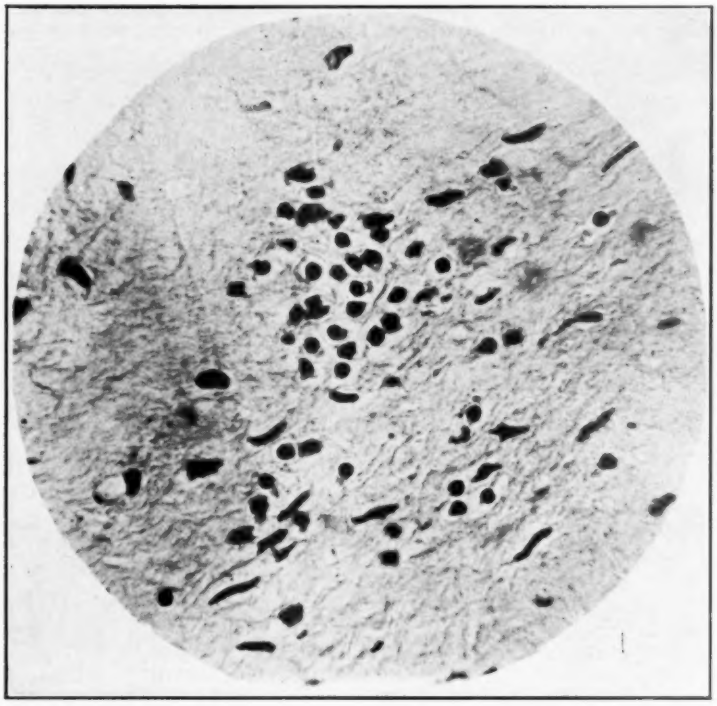

Fig. 2.—Photomicrograph of small lymphocytic focus in the substance of a mitral valve, illustrating the comparatively slight evidence of inflammation which placed this case in the *intermediate group*.

Table I represents the eliminated hearts containing arteriae valvulares, together with data concerning their sites and forms as well as reasons for eliminating them from this series.

Table II represents a brief résumé of the statistics of the 123 hearts possessing arteriae valvulares out of the 700 studied. These statistics

TABLE I

NUMBER OF HEART	TYPE OF VASCULATURE IN AORTIC LEAFLET OF MITRAL VALVE	REASON FOR ELIMINATION FROM NORMAL SERIES
5268	Incomplete form	Small foci of lymphocytes in mitral valve
5335	Incomplete form	Small foci of lymphocytes in mitral valve
5494	Incomplete form	Small foci of lymphocytes in mitral valve
X-349	Incomplete form	Bacterial embolus in vessel in valve (probably agonal)
5716	Incomplete form	Foci of lymphocytes in mitral valve
5661	Incomplete form	Pericarditis, terminal
5449	Incomplete form	Peculiar inflammatory lesion in left auricle
5524	Incomplete form	Cellular foci in myocardium
5738	Incomplete form	Lymphocytes and plasma cells in mitral valve
5751	Incomplete form	Small focus of lymphocytes in mitral valve
5761	Incomplete form	Pericarditis, terminal. Few lymphocytes in mitral valve
5609	Incomplete form	Bacterial emboli in posterior papillary muscle and cellular infiltration in right ventricle
5491	Blood vessel in tricuspid valve; incomplete form	Fusion of cusps of aortic valve. History of scarlet fever
5715	Complete form	Diffuse lymphocytosis in mitral valve
5538	Complete form	Lymphocytes in myocardium and valve
S244	Complete form	Capillaries resembling granulation-tissue type but no inflammation
5297	Complete form	Small perivascular focus of lymphocytes in mitral valve

TABLE II

	WITH COMPLETE MITRAL VASCULARIZATION	WITH INCOMPLETE MITRAL VASCULARIZATION	TOTAL
Hearts with endocarditis, valvulitis, or other inflammatory focus	48	61	109
Normal hearts (in which no cause can be found to which the existence of the arteriae valvulares can be attributed)	6	8	14
			123

TABLE III

	WITH COMPLETE MITRAL VASCULARIZATION	WITH INCOMPLETE MITRAL VASCULARIZATION	TOTAL
Hearts with endocarditis or valvulitis	44	48	92
Normal hearts (in which no cause can be found to which the existence of the arteriae valvulares can be attributed)	10	21	31
			123

are based on the acceptance of the 14 hearts only as being normal and at the same time containing vascularized valves.

Table III represents the same statistics as those in Table II, but here the *intermediate group* has been included in the series of hearts with

arteriae valvulares, but without endocarditis. The purpose of including this *intermediate group* lies in the fact that this may represent a closer approximation of the number of hearts found in the autopsy material studied which show vascularized valves but in which no definite cause can be found to account for the vascularization.

ABSTRACTED PROTOCOLS

In the protocols which follow we made efforts to obtain histories of some definite etiological factor which might suggest the occurrence of an endocarditis in the past. With the exception of one patient who had a past history of scarlet fever, in none of these cases was there obtained a positive history of rheumatic fever, chorea, scarlet fever, arthritis, repeated attacks of tonsillitis, or other specific infectious diseases. In some cases, however, no specific reference was found in the hospital records concerning the occurrence of these diseases in the past.

CASE 1.—(Mount Sinai 5457) Patient M. F., female, fifty-three years of age, was admitted to the Mount Sinai Hospital with signs and symptoms of acute myocardial insufficiency on an atherosclerotic basis. The patient did not yield to stimulation and died several hours after admission.

Summary of Autopsy Findings: Slight atherosclerosis of coronary arteries; chronic interstitial pneumonia; chronic adhesive pleuritis; chronic emphysema and bronchitis; cholelithiasis. The heart weighs 500 gm. The right heart is definitely hypertrophied. The tricuspid valve shows a diffuse atherosclerotic thickening with ridge-like formation (tension type) of the closing edge. Otherwise the heart shows no gross abnormalities. The aortic cusp of the mitral valve is the seat of vascularization of the incomplete type. Microscopy of the heart: Moderate hypertrophy of fibers of the left posterior papillary muscle. Hypertrophy of muscle fibers with patchy perivascular fibrosis in right ventricle. The aortic cusp of the mitral valve shows blood vessels of the arterial type as well as muscle bundles. There are no inflammatory foci to be seen.

CASE 2.—(Mount Sinai 5508) Patient C. O., female, fifty-seven years of age, was admitted to the Mount Sinai Hospital with signs and symptoms of a carcinoma of the stomach with visceral metastases. Died 17 days after admission.

Summary of Autopsy Findings: Medullary carcinoma of the stomach with metastases in the liver and regional lymph nodes. The heart is grossly negative except for slight atherosclerotic thickenings of the mitral and aortic valves. The aortic cusp of the mitral valve is the seat of a vascularization of the incomplete type. Microscopy of the heart: The left ventricle shows hypertrophied muscle fibers, irregular diffuse fibrosis and arteriosclerosis of the blood vessels. The aortic cusp of the mitral valve shows some atherosclerosis and possesses blood vessels of the arterial type as well as muscle bundles. There are no inflammatory foci to be seen.

CASE 3.—(Mount Sinai 5509) Patient W. O., female, twenty-one years of age, was admitted to the Mount Sinai Hospital in coma. Patient died several hours after admission.

Summary of Autopsy Findings: Marked hyperplasia of Peyer's patches and solitary lymph follicles of the intestines. Cloudy swelling of organs. Diagnosis:

Early typhoid fever. Grossly the heart is normal. The aortic cusp of the mitral valve is the seat of a vasculature of the incomplete type. Microscopy of the heart: The vessels of the posterior papillary muscle show a moderate degree of arteriosclerosis. Other sections through the myocardium are negative. The mitral valve possesses arterial blood vessels and muscle bundles. There are no inflammatory foci to be seen.

CASE 4.—(Mount Sinai 5565) Patient J. S., female, fifty-four years of age, was admitted to the Mount Sinai Hospital in collapse. On account of the past history, a diagnosis was made of a perforated peptic ulcer. Patient died one hour after admission to the hospital.

Summary of Autopsy Findings: Acute hemorrhagic pancreatitis. The heart shows no gross abnormalities. The aortic cusp of the mitral valve is the seat of a vasculature of the incomplete type. Microscopy of the heart: Sections through the myocardium on the left side show a mild hypertrophy. The mitral valve possesses arterial blood vessels superficially situated, and muscle bundles. There are no inflammatory foci to be seen.

CASE 5.—(Mount Sinai 5566) Patient, J. W., male, forty-six years of age, was admitted to the Mount Sinai Hospital with signs and symptoms of a perforated duodenal ulcer. Laparotomy was performed, and the patient died the following day.

Summary of Autopsy Findings: Perforated duodenal ulcer with acute generalized peritonitis; cavernoma of the liver; atherosclerotic thickening of the main branches of the coronary arteries. The heart is slightly atrophic. The tricuspid and aortic valves show slight atherosclerotic thickenings. The aortic cusp of the mitral valve is the seat of a vasculature of the incomplete type. Microscopy of the heart: The left auricular and ventricular musculatures show focal scarring. The blood vessels show marked endarteritic and atherosclerotic changes. The aortic cusp of the mitral valve shows blood vessels of the capillary type superficially placed, as well as muscle bundles. There are no foci of inflammation to be seen.

CASE 6.—(Mount Sinai 5570) Patient B. T., female, twenty-nine years of age, was admitted to the Mount Sinai Hospital with signs and symptoms of thrombocytopenic purpura. A splenectomy and a transfusion were performed. Patient died shortly after the operation.

Summary of Autopsy Findings: Subdural and subarachnoidal hemorrhages; hemorrhages in the left suprarenal gland and in most of the other organs. The heart is normal in size and shape. The pericardium shows many purpuric spots. The mitral and aortic valves show slight tension thickenings. The heart is otherwise grossly normal. The aortic cusp of the mitral valve is the seat of a vascularization of the incomplete type. Microscopy of the heart: Sections of myocardium are negative. The aortic cusp of the mitral valve contains blood vessels of the arterial, venous, and capillary types as well as muscle bundles. There are no inflammatory foci to be seen.

CASE 7.—(Mount Sinai 5597) Patient N. S., male, sixty-six years of age, was admitted to the Mount Sinai Hospital with signs and symptoms of arteriosclerotic gangrene of the left foot. Amputation of the left foot was performed. The patient gradually declined and died six days after admission.

Summary of Autopsy Findings: Atherosclerosis of coronary arteries with marked narrowing of anterior descending and circumflex branches of the left coronary artery. Generalized atherosclerosis. The heart presents no gross abnormalities with the exception of the vascular disease mentioned. The aortic cusp of the mitral valve is the seat of a vascularization of the incomplete type. The posterior portion of the aortic cusp of the mitral valve as well as the posterior cusp of the mitral valve

shows thickening of the edge. Microscopy of the heart: The left posterior papillary muscle shows focal scarring. The blood vessels in the mitral valve were of the arterial and capillary types. Muscle bundles are present. The tips of the valve flaps show marked atheromatous changes but no inflammatory reactions are found.

CASE 8.—(Mount Sinai 5627) Patient M. S., female, thirty-six years of age, was admitted to the Mount Sinai Hospital complaining of abdominal pain, distention and vomiting for two days following an instrumental labor. The patient rapidly went into a state of collapse and died the day following her admission.

Summary of Autopsy Findings: Laceration of cervix uteri and the right broad ligament; hemoperitoneum; collapse of lower lobe of left lung. The heart is grossly negative. The aortic cusp of the mitral valve is the seat of a vascularization of the incomplete type. Microscopy of the heart: The left ventricle, the left posterior papillary muscle, the interventricular septum and the right ventricle show focal fibrosis and arteriosclerosis of the coronary branches. The aortic cusp of the mitral valve contains arterial vessels, capillaries, and muscle bundles. There are no foci of inflammation.

CASE 9.—(Mount Sinai 5665) Patient, M. H., male, forty-five years of age, was admitted to the Mount Sinai Hospital with signs and symptoms of a gastrojejunal ulcer (he had previously had a gastroenterostomy performed). A subtotal gastrectomy was performed and two days later the patient died with signs of bronchopneumonia.

Summary of Autopsy Findings: Bronchopneumonia and diffuse edema and congestion of the lungs; slight parenchymatous degeneration of the kidneys. The heart is grossly normal in size and shape. The aortic and mitral valves show some evidence of atherosclerotic thickening. The mitral valve is the seat of a vasculature of the complete type. Microscopy of the heart: The coronary arteries and their branches show arteriosclerotic changes. The mitral valve contains numerous vessels of the arterial, venous, and capillary types. There are no inflammatory foci.

CASE 10.—(Mount Sinai 5743) Patient P. S., male, thirty-one years of age, was admitted to the Mount Sinai Hospital with signs and symptoms of lobar pneumonia. On the tenth day after admission the patient sank into a typhoidal state and died on the fifteenth day of his illness.

Summary of Autopsy Findings: Lobar pneumonia in various stages of consolidation in all lobes; acute fibrinous and chronic adhesive pleuritis on the left side; acute fibrinous pericarditis; right renal calculi and hydronephrosis. The heart is normal in size and shape. The aortic cusp of the mitral valve is the seat of a vasculature of the complete type. Microscopy of the heart: The mitral valve shows early atherosclerotic changes, a number of blood vessels superficially situated, and muscle bundles penetrating the valve substance. No inflammatory foci are seen.

CASE 11.—(McGill 55) Patient Y. C., female, twenty-nine years of age, was admitted to the Royal Victoria Hospital, Montreal, with typical signs and symptoms of typhoid fever. Two weeks after admission she had a hemorrhage from the bowel and soon after died.

Summary of Autopsy Findings: Typhoid ulcerations in the ileum with focal necroses in the mesenteric glands and in the spleen. Bronchopneumonia with infarction in the right lower lobe of the lung. Chronic adhesive pleuritis and chronic fibrous adhesions in the pelvic peritoneum. The heart is grossly normal and weighs 250 gm. A vascular structure of the complete type is seen in the aortic cusp of the mitral valve. Microscopy of the heart: Several sections through the heart muscle show moderate focal fibrosis. Aortic cusp of the mitral valve shows numerous

large vessels of the arterial, venous, and capillary types. The latter are seen in abundance in the region of the line of closure. Many of the arteries show atherosclerotic changes and in places, obliteration. Scattered muscle bundles are found in the valve. No inflammatory foci are seen.

CASE 12.—(Yale 1113) Patient, P. O'C., male, forty-six years of age, was admitted to the New Haven General Hospital with typical signs and symptoms of cardiorenal failure. Soon after admission the patient developed signs of bronchopneumonia and pulmonary edema. He died five days after admission.

Summary of Autopsy Findings: Chronic glomerulonephritis, cardiac dilatation, and hypertrophy with the usual findings of congestive heart failure. Central necrosis of the liver; organizing mediastinitis; pleuropericarditis. The subsidiary findings are chronic apical tuberculosis; healed duodenal ulcer; nephrolithiasis; hydro-

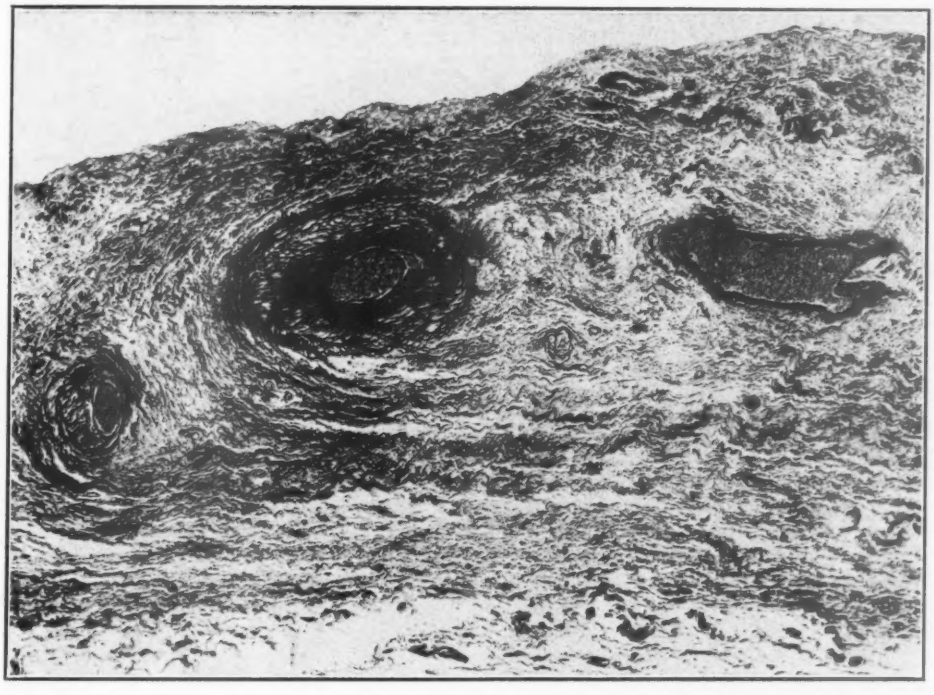

Fig. 3.—Photomicrograph (high power) of arteriosclerotic blood vessels in the aortic cusp of the mitral valve from a normal human heart. Note the arteries, veins, and capillaries.

nephrosis with cysts of the kidney. The heart is concentrically hypertrophied and also dilated. It weighs 675 gm. There is no evidence of valvular disease. The aortic cusp of the mitral valve shows a typical vascularization of the complete type. Microscopy of the heart: Sections through various portions of myocardium show hypertrophied musculature, focal fibrotic areas, and arteriosclerotic changes in the coronary arteries and branches. The mitral valve shows numerous vessels of the arterial, venous, and capillary types as well as muscle bundles. The arterial vessels show hypertrophy, arteriosclerotic changes, and narrowing (Fig. 3). There is no evidence of inflammation. The aortic valve shows intense arteriosclerosis.

CASE 13.—(Yale 1203) Patient A. S., male, sixty-six years of age, was admitted to the New Haven General Hospital with signs and symptoms of a carcinoma of the stomach. At operation the pyloric antrum and a portion of the head of the

pancreas were excised. There was marked hemorrhage, and the patient died in shock soon after the operation.

Summary of Autopsy Findings: Hemoperitoneum; chronic adhesive pleuritis; adenoma of the thyroid. Specimen removed at operation showed an adenocarcinoma of the pyloric antrum. The heart weighs 350 gm. and shows no gross abnormalities. The aortic cusp of the mitral valve shows a vasculature of the complete type. Microscopy of the heart: Several sections from the myocardium of the left side show a number of focal areas of fibrosis with atherosclerotic changes in the coronary artery branches. The aortic cusp of the mitral valve shows atheromatous changes, arterial and venous vessels, and muscle bundles. The arteriae valvulares show atherosclerosis. There are no signs of inflammation.

CASE 14.—(Yale 1276) Patient E. B., male, sixty-two years of age, was admitted to the New Haven General Hospital with a right hemiplegia. During his stay in the hospital a right bundle-branch block developed. Patient died with a terminal pneumonia thirty-four days after admission.

Summary of Autopsy Findings: Patchy bronchopneumonia involving all lobes of the lungs; several acute ulcers near the pylorus of the stomach; hypertrophic arthritis of spinal vertebrae. The heart weighs 600 gm. and shows hypertrophy and dilatation of both sides. There is no evidence of valvular disease. The aortic cusp of the mitral valve shows vasculature of the complete type. Microscopy of the heart: There are small scattered areas of fibrosis in the myocardium. The coronary arteries, particularly on the left side, show atherosclerotic changes. The mitral valve shows vessels of the arterial and capillary types but no signs of inflammation.

DISCUSSION

A review of the 14 cases abstracted above shows that we are dealing with blood vessels in the valves of hearts from patients where no clinical or appreciable pathological evidence exists to suggest that these vessels may have arisen on the basis of an inflammatory lesion in the valves. They can therefore be used as strong additional evidence to bear out our contention as to the existence of noninflammatory blood vessels in some valves of isolated human hearts.

Several questions may still be raised as to the validity of this evidence. We shall present the questions together with our comments on them:

1. Is it possible that these valves had really been the seat of an endocarditis, that granulation tissue blood vessels had in this way been produced, and that all signs of inflammation in the valve had disappeared without leaving appreciable traces other than the blood vessels?

The valves examined in this series showed occasionally such thickenings at the closure line as were consistent with the age of the individual and were similar to those found so often in valves of perfectly normal hearts. These "tension changes," as they are called by Dr. Libman, taken by themselves are insufficient evidence of the preexistence of inflammation, although it must be admitted that it is often very difficult, if not impossible, to distinguish histologically between a healed endocarditis and a so-called "wear and tear" thickening.

Some of the valves, however, did not show any appreciable thickening. Moreover, the main portions of the blood vessels in all cases lay within perfectly intact valve structure, and we know of no instance where an inflammatory lesion has come to complete restitution of integrity, leaving only intact blood vessels in its path. If anything, organizing granulation tissue should lead to compression and disappearance of blood vessels rather than to their persistence. Taken all in all, however, this histological evidence, alone, cannot be used as conclusive proof that no inflammation preceded the existence of the vessels.

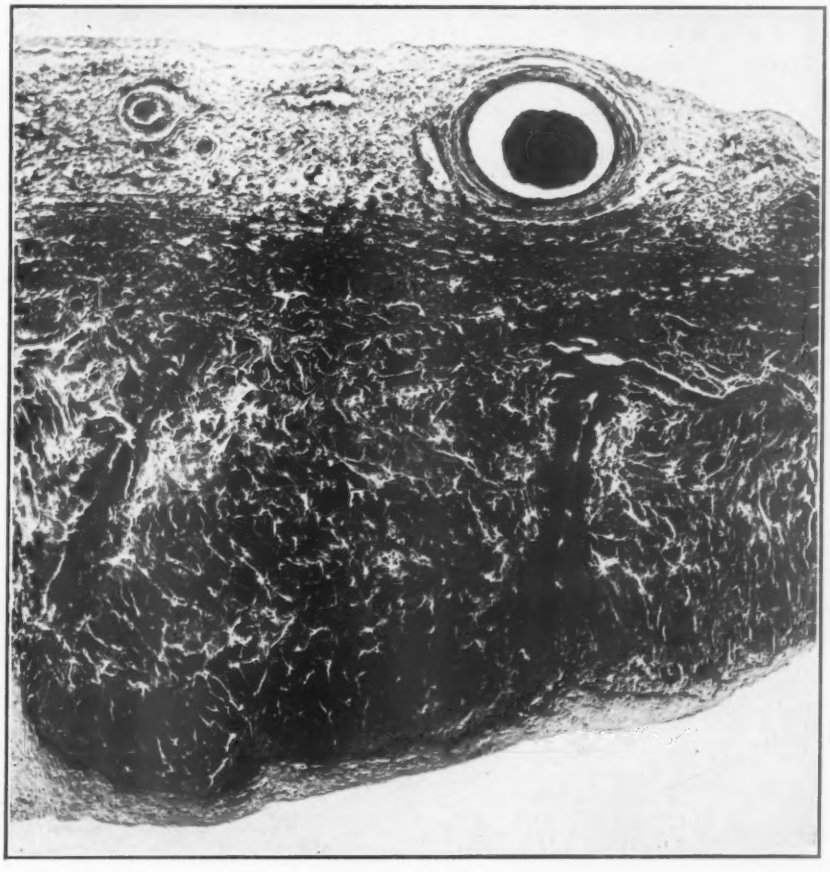

Fig. 4.—Photomicrograph (high power) of blood vessels (arteries, veins, and capillaries) in the aortic cusp of the mitral valve from a normal human heart.

2. Is it possible that even though many of the blood vessels which we find in these valves are distinctly of the arterial and venous types (Fig. 4), they may none the less have arisen as granulation-tissue vessels?

For example, it has been suggested that vessels, such as those found in the condition of the eye known as pannus, are newly formed and may possibly possess muscular and elastic walls. We may say to this that we have been unable to find in the literature accurate data

bearing out this contention nor have we seen in numerous preparations of such specimens, histologically and anatomically distinct arteries and veins. Again, it has been contended that the recanalization of a thrombosed vessel at times presents newly developed muscular and elastic walls in the lumen of the old vessel. This we have seen, and while we cannot deny the possibility of its occurrence in the blood vessels in the valves, in order to make such a process responsible for the structure of these vessels, it forces us to assume that valvulitis, almost to the total exclusion of all other diseases, is peculiarly disposed toward new formation of elastic and muscular walls within the capillaries of its granulation tissue. Furthermore, we have found no evidence of thrombosis in these vessels.

3. Is it possible that since 109 out of 123 hearts with arteriae valvulares were associated with valvulitis (Table II) an endocardial lesion was overlooked in the 14 cases presented above? For example, whereas we were unable to find evidence of a healed endocarditis in our microscopical sections, had we made serial sections of the complete valves, might we have found an endocardial focus of inflammation?

In answer to this question we wish to point out that the occurrence of arteriae valvulares in such valves as could hardly have been the seat of inflammatory vascularization is by no means rare. For if we consider Table III which shows the statistics including the *intermediate group* of cases (i.e., those cases which have been eliminated on account of extravalvular lesions, small lymphocytic foci in the valves, etc.), we find that 25 per cent of hearts (31 out of 123) with vascularized valves showed no appreciable evidence of a preexisting endocarditis. If we accept only the 14 hearts described here as being normal, we still have 11.5 per cent (14 out of 123) of this group or 2 per cent (14 out of 700) of 700 consecutive autopsies showing valvular blood vessels without appreciable evidence of antecedent inflammatory disease.

Furthermore, although we have not serially sectioned the valves of the hearts recorded here, we have taken numerous sections through the valves, particularly at the sites of distribution of the vessels. If these vessels were truly of an inflammatory nature some evidence of inflammation, past or present, should have been found in their proximity unless, as we stated before, we assume that all appreciable traces of inflammation had disappeared in these hearts. We shall see that this alternative leads us to conclusions difficult to accept. Moreover, the fact that the arteriae valvulares do not spring from neighboring capillaries but from the arteriae anastomotica auricularis magna which at times is situated fairly high in the auricular septal musculature, the fact that vestiges of these vessels are almost invariably found in normal hearts without arteriae valvulares, the fact that

gradations exist between the complete type in aborted form (i.e., within the auricular musculature at the base of the mitral valve) and the form in which the arcuate arch is near to the closure line of the valve flap, and the fact that the vessels in the incomplete types are found at the same sites as the arteriae valvulares in the complete types, make it very difficult to accept these vessels as of granulation tissue origin.

An important argument in support of the noninflammatory nature of these vessels is the fact that they are found with remarkable frequency in such animals as pigs, sheep, and cattle. Moreover, in these animals, as we have pointed out in a previous publication, the architecture of these vessels bears a striking resemblance to that found in the human heart. It would seem, therefore, that a large percentage of pigs, sheep, and cattle develop valvulitis immediately after birth, that the resemblance between the human and animal structures is more apparent than real, or that the vessels in the human heart are indeed homologous to those found so frequently in these animals. There seem to be no good reasons for accepting the first two of these possible conclusions.

We are quite prepared to believe that a vascularized valve which becomes the seat of an inflammation may develop a secondary capillary vasculature of granulation-tissue type, but if we assume that the arteriae valvulares are also of granulation-tissue origin, we are confronted with difficulties which appear much greater than those cited against their acceptance as of this nature.

We do not need to reiterate in detail the observations favorable to considering these vessels as of embryonic derivation, viz., their structure, source, architecture, analogy to the aborted type in practically all human hearts, and to the arteriae valvulares seen in certain animals. On the other hand, if it be assumed that in the 14 cases reported here the patients had suffered from an endocarditis in such severe form as to produce fully formed blood vessels, we must also assume that at least 2 per cent of the 700 individuals who came to autopsy in the hospitals from which this material was obtained (possibly 4 per cent if we accept the *intermediate group*) developed during the course of their lives a severe valvulitis or endocarditis which is of unknown etiology, of unknown type, producing no appreciable clinical manifestations such as we know of other forms of endocarditis and healing without leaving any appreciable traces other than fully formed blood vessels. We feel at present that we are not prepared to accept this alternative.

SUMMARY

Seven hundred human hearts have been studied with reference to the existence of blood vessels in the valves. Of these, 14 normal hearts have been described which present blood vessels in some of the valves.

A thorough clinical and pathological examination has failed to reveal that these vessels owe their origin to inflammation. Additional reasons are given which render it highly improbable that they owe their origin to inflammation, or that they are other than embryonic vestiges.

We wish to express our thanks to Drs. Horst Oertel, M. C. Winternitz, and R. G. Hussey for having furnished us with some of the material used in this study, and to Mrs. Eric Frisch for valuable technical assistance.

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PULSATING SPLEEN IN AORTIC INSUFFICIENCY

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IN THE *Observationes Medicae* of Nicolas Tulp,*[1] there is reported an extraordinary case of pulsating spleen under the name of lien verberens.

"Nothing in medical art is better known than that the spleen pulsates continually, if violently moved by the arteries. But for this organ itself to strike the ribs so forcibly that the sound of the beating (lit. flogging, or whipping) may be heard from afar, that certainly is novel, and perchance hitherto unheard of.

"In the case of Nicolaus Fabrus, an active man, but rather frequently afflicted with black bile, a hardened (indurated) spleen made so forceful an impulse on the adjacent ribs, that not only he himself felt pain therefrom, but persons at a considerable distance might clearly hear the loud sound of the beats; and even so distinctly that one might count the separate impulses, and with the close-pressed hand feel the throb of the beating spleen.

"In fact, I remember that in company with Henricus Salius, the physician of Utrecht (?), I heard these repeated sounds at a distance of above thirty paces. But according as the black bile overflowed more profusely or more scantily, the vehemence of the beats persisted or abated. As soon as the body was agreeably emptied, sometimes these pulsations ceased entirely, to reappear when the bile again became impure, so that he was forced repeatedly to seek medicinal aid against the otherwise unmitigable complaint.

"But whence this repetious series of beats? Is it not from the movement of the heart, whose systole and diastole are communicated, as in the entire body, forcibly to those arteries which are so numerous about the spleen that Adrianus Spigelius, the energetic Hercules of anatomic study, does not hesitate to state them to be four times as numerous as veins? (Fabric, human., Bk. iv, Ch. xiii.)

"But what produced the sound? Was it not the hard texture of the spleen, for a hard substance resists? As long as it is soft, like a sponge, it gives forth no sound. The surface of the spleen hardened, and smote against the ribs in its repeated movements. (Describes a spleen 'incrusted with hard cartilage.')"

A considerable time elapsed before another case was observed and recorded. This was reported by C. Gerhardt.[2] The patient, a white man, twenty-seven years old, and a smith by occupation, had aortic insufficiency and a tertiary intermittent fever. The spleen pulsated, and as it could be grasped in the hand, it was evident that the pulsation was expansile. It was not a transmitted pulsation from the heart. Over the tumor was heard a dull, double note, and only at the upper border were the heart sounds audible. There was no notable increase in the leucocytes. Large doses of quinine finally cured the patient, and the tumor was also influenced by faradization.

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*Nicolas Tulp, Physician and Magistrate of Amsterdam. Born Oct. 11, 1594; died Sept. 12, 1674. He celebrated the fiftieth anniversary of his election as alderman in 1672. His portrait appears as the teacher in the "Lesson in Anatomy" by Rembrandt. He published *Observationes Medicae* of which the first edition appeared in 1641, and the fifth in 1716.

Gerhardt also observed two other cases in patients with well-compensated aortic insufficiency, both with pericarditis and fever. In all three it was a soft swelling similar to that felt in a pulsating jugular vein. Gerhardt regards this as a characteristic sign caused by the abnormal blood-pressure conditions of aortic insufficiency, and the relaxation of the blood vessels produced by the fever. It is produced by conditions similar to those that cause the capillary pulse. It is, therefore, another vascular sign occurring in aortic insufficiency.

Five year later J. Prior[3] reported two new cases. A man thirty-seven years old, who had had aortic insufficiency for several years, developed typhoid fever. During the stadium an expansile pulsating spleen could be felt. The pulsation showed a definite relation to the force of the heart and became weaker as the heartbeat also weakened. When the patient finally recovered, the splenic pulsation could no longer be felt even though there was some disturbance of compensation. The second case, a man of thirty-seven years, without murmurs, but with a hypertrophied heart, visible pulsation of the peripheral arteries, and a pulse tracing similar to that of aortic insufficiency, had a croupous pneumonia, during the course of which the spleen enlarged and showed an expansile pulsation. This disappeared as the patient recovered by lysis. Prior agrees with Gerhardt that the association of aortic insufficiency and a febrile condition is required to produce the pulsation. He also mentions that Maissurianz (*St. Petersburgh Med. Wchnschr.*, 1884, lii, 517) noted a systolic murmur over an enlarged spleen.

During the past summer (1927) there was admitted on the service of my colleague, Dr. A. A. Stevens, a patient whom I studied, and it is by his courtesy that I am permitted to report this case.

J. D., aged twenty-seven years, white, was admitted to the Philadelphia General Hospital July 10, 1927 and died August 8, 1927. On admission his chief complaint was of some disturbance of the heart. He stated that in February, 1927 he had begun to feel giddy. The condition was somewhat variable; he felt well for a week, and then became restless. He had no pain, but he felt so tired that he wanted to lie down all the time. He stopped work in February, and had been up and down since then until two weeks prior to admission, when it was noted by the district nurse that he had fever, and he was kept in bed. He stated that if he lay on the right side, he had pain in the right shoulder, really more of a soreness than of pain. He felt stronger, but was tired of lying in bed. He always felt his heart beating. He had no dyspnea, but had swelling of the legs when out of bed; the swelling disappeared if he remained in bed. He had had no cold, and no sore throat. His appetite had been fair, but the thought of certain foods nauseated him. There had been much eructation of gas and chronic constipation, for which he took pills. He had lost weight, but he did not know how much. He had nocturia three or four times, but no frequency during the day, and no dysuria.

Previous Medical History: He had never had measles, pertussis, chicken-pox, scarlet fever, pneumonia, malaria, influenza, tonsillitis, chorea, or venereal disease. Ten years before, when he was sixteen years old, he had an attack of articular,

migrating rheumatism, and he was in bed for three or four months. After recovery, he felt well until the onset of the present condition. In the interval he never had dyspnea or edema of the legs. He had broken his right elbow, and had had three or four operations upon it subsequently, without complete restoration of the function. He suspected some trouble with his teeth. On more careful questioning he admitted that he had had attacks of sore throat and some growing pains when he was a child.

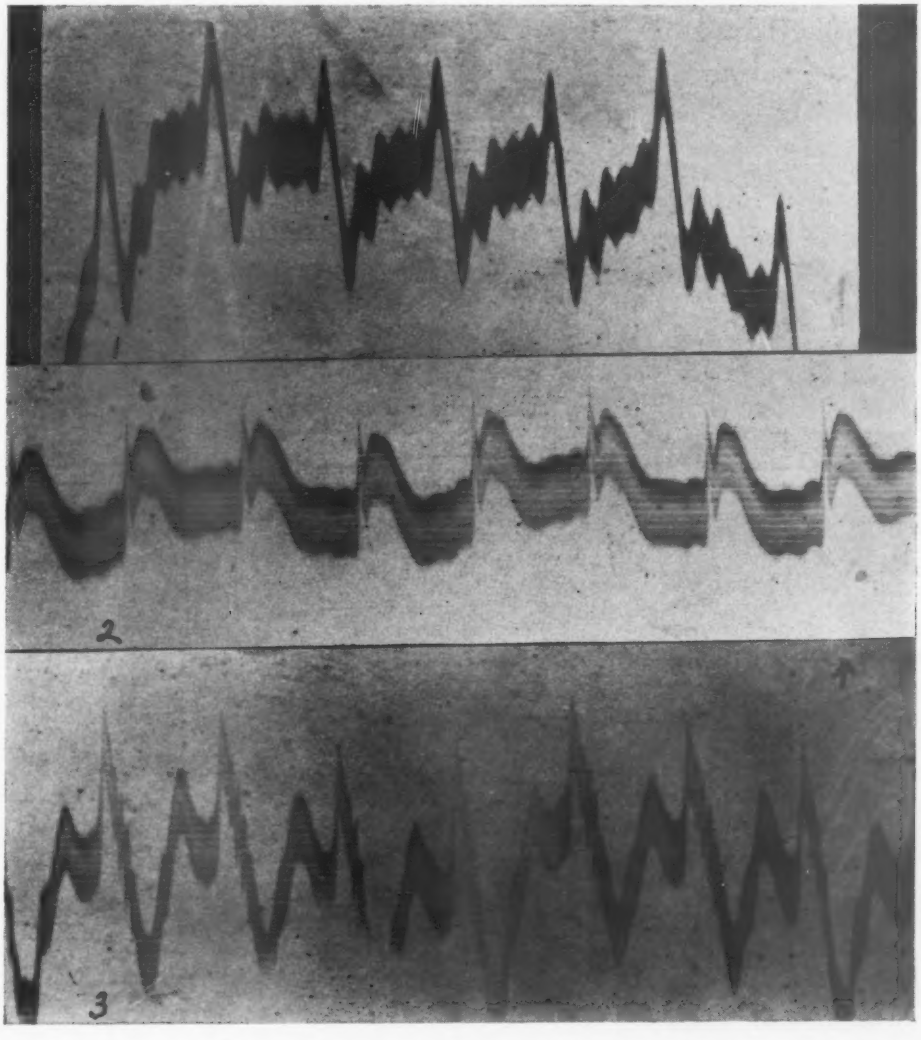

Fig. 1.—Tracings from the case of pulsating spleen, taken by reflecting a beam of light from a Franck mirror or capsule. *1*, Tracing from the pulsating spleen, showing a series of small waves which are not found in the tracings from the abdominal aorta or from the carotid artery, and which are therefore peculiar to the pulsating spleen. *2*, tracing from the abdominal aorta. *3*, Tracing from the carotid artery.

Family History: His father was living and well. His mother died of a stroke at the age of fifty-five. One sister and two brothers were living and well; two brothers died in early childhood. There is no history of tuberculosis, cancer, or insanity in the family.

His occupation was that of a mechanic. He smoked cigarettes very moderately, and used alcohol very little.

Physical Examination: He was thin and emaciated, and appeared to be very sick. His pulse was of the Corrigan type. His blood pressure in the right arm was 130/42 mm., the final phase being zero; in the right leg it was 152/42 mm., the last phase being zero. DeMusset's sign was present. The pupils were dilated and reacted to both light and accommodation. The ocular motions were normal. The ears and nose were normal. The mouth showed gingivitis and slight pyorrhea. The tongue was moist and slightly coated. The throat was clean. There was a marked pulsation in the neck, with a thrill on pressure over the subclavians. There were a few palpable cervical glands. The chest was of the long, slender type; the expansion equal. Tactile fremitus was normal throughout, except at the left apex posteriorly, where it was slightly impaired. The breath-sounds were vesicular, but slightly diminished over the left apex posteriorly. No râles could be heard. There was no bronchophony or pectoriloquy, but the whispered voice was heard with increased intensity over the entire chest. Over the cardiac area a marked precordial heaving could be observed. There was an intense systolic and a fainter diastolic thrill. The left border of the heart was 15 cm. to the left of the median line, in the fifth interspace. The right border was 4 cm. to the right in the fourth interspace. In the second interspace the dullness was 8 cm. broad. At the apex the heart-sounds were loud, and there was a loud systolic murmur with a short, moderate diastolic murmur. In the fourth interspace there was a loud, metallic second sound, followed by a short diastolic murmur. At the base there was a faint systolic murmur, followed by a loud diastolic murmur that was transmitted down the sternum. The aortic second sound was louder than the pulmonic second sound. The abdomen was flat. The liver extended below the costal margin. The kidneys were not palpable. The right elbow was deformed; the head of the radius was enlarged, and the joint could not be fully straightened. The fingers were clubbed. There was a capillary pulse; no edema of the legs. The knee-jerks were active.

At the time of admission the compensation seemed to be fair. I suspected blood-stream infection with the Streptococcus viridans, because of the peculiar grayish pallor of the patient's skin.

On July 13 the patient had some toothache and diarrhea. The spleen could be palpated about 2 cm. beneath the costal margin, and it was slightly tender. The patient ascribed the tenderness to having taken a dose of salts.

On July 15 the spleen was still palpable, but no longer tender. No petechiae could be found.

On July 16 it was noted that the spleen was pulsating distinctly. The pulsation was synchronous with the heart and seemed to be expansile. This pulsation was noted subsequently nearly every day, and increased moderately until the *first of August.*

On August 1 a positive blood culture of the Streptococcus viridans was reported.

On August 3 the patient was distinctly weaker; the face was slightly swollen; the hands were cyanosed; he had a slight sore throat, and the spleen seemed to be pulsating less vigorously.

On August 4 the whole right side of the face was swollen. He steadily grew worse, developed dyspnea, which was relieved by morphine, and finally died on August 8. Transfusion had been considered, but it was impossible to obtain a donor. No autopsy was performed.

The following laboratory reports may be included:

The urine varied in specific gravity from 1.022 to 1.028; the amount of albumin from a trace to a cloud; there was no sugar; no casts could be found; there were a few white blood cells; no red blood cells, but a great excess of mucus.

On July 12 the red blood cells were 3,000,000; the white blood cells 11,200; and the hemoglobin 84 per cent. The differential count gave polymorphonuclears 95; lymphocytes, 4; transitionals, 1. Anisocytosis and poikilocytosis were present.

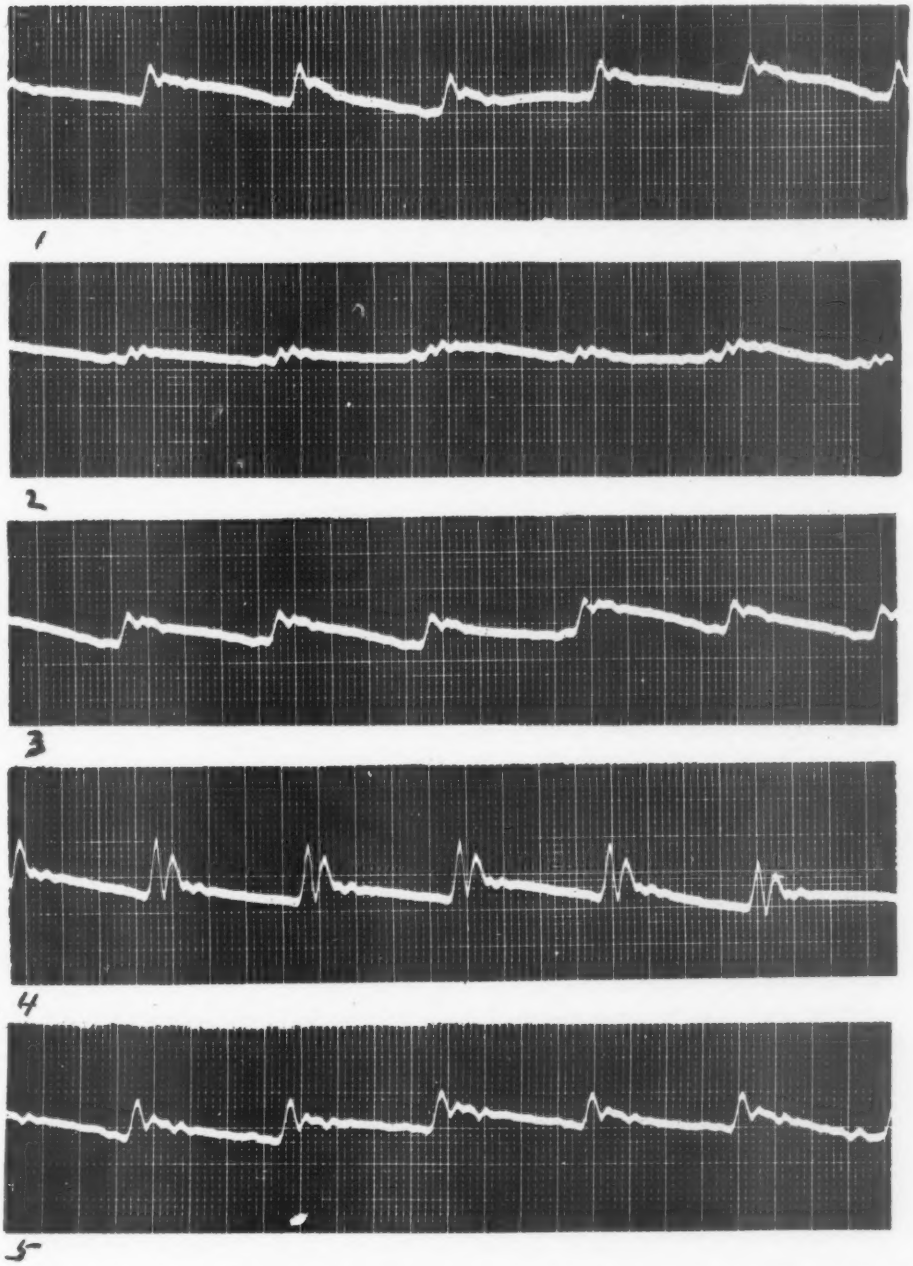

Fig. 2.—Tracings to show the pulsation of an abdominal tumor. These tracings were obtained by the usual method of using a tambour attached to a delicate lever, which vibrated in front of the ray of light of a string galvanometer. The tracings from over the tumor are essentially the same as those of the aorta and the carotid artery, although the amplitude of the waves in the tracing of the aorta is greater. *1*, taken from the point of maximum pulsation of the tumor; *2*, taken from some distance to the left; *3*, taken from a slight distance to the right; *4*, pulsation of the abdominal aorta just above the umbilicus; *5*, pulsation of the carotid artery.

The Wassermann was negative. Blood chemistry was normal. The blood was found to be Type III. A blood culture was taken, which was reported still sterile on July 20. On July 29 the blood culture was taken, which was reported on August 1 as positive for the Streptococcus viridans. At this time the serum agglutinated the streptococcus cardioarthritidis in a solution of 1:80.

On August 4 the red blood cells were 3,150,000.

It was my belief, when I read Litten's[4] reference to pulsation of the spleen many years ago, that the condition would be found with reasonable frequency, but since then I have observed two doubtful cases, and only one that conformed to the others reported. Moreover, a cursory search through some score of works on heart disease reveals no mention of it, although Mackenzie (*Diseases of the Heart*, ed. 3, London, 1913, p. 340) mentions a movement of the liver, produced by transmitting the movement of the heart and resembling pulsation. Gerhardt mentions two additional cases, but does not describe them.

If one can draw conclusions from four cases described in detail, the conditions necessary to cause the pulsation of the spleen are a severe type of aortic regurgitation and an infectious process. In the three cases reported the infections have been malaria, doubtful because diagnosed before the plasmodium had been discovered, typhoid fever, pneumonia, and streptococcic blood-stream infection, probably associated with endocarditis. Gerhardt suggests that the infection relaxes the splenic blood vessels, but this seems to me to be inadequate. The enlargement and softening of the spleen that occurs in the infection, however, must contribute to the occurrence, the chief cause of which is the excessive pulse pressure that occurs in aortic regurgitation. Gerhardt also mentions the change in the intensity of the pulsation that parallels the apparent variation in the force of the heartbeat; and this was manifest in the case that I report. The case of Tulp is interesting because, if I am not mistaken, he is the first to report the pistol-shot sound of Traube, at least so I interpret the loud sound that he described and that he supposed to be caused by the beating of the spleen against the ribs.

Pulsation of the spleen, owing to its rarity, can be of no great importance, and even if it were more common, would probably prove to be no more than an additional sign of aortic insufficiency, for it is doubtful whether or not it can occur in any other condition. Nevertheless, it is interesting to observe.

It so happened that there was admitted to my service at the Presbyterian Hospital, in the men's medical ward, a patient who had a pulsating tumor of the epigastrium. This tumor was apparently growing in the wall of the stomach near the pylorus, and the pulsation seemed to be transmitted. Dr. McMillan again was kind enough to take tracings of this pulsation. They show a very different picture from those obtained from the pulsating spleen. Three tracings were

taken from the tumor, one at the point of maximum pulsation, and two slightly distant from this; and they differ only in degree. There were also two tracings taken, one from the abdominal aorta, just beneath the pulsating mass, and the other from the carotid. They show very clearly that the pulsation is in all respects similar to the pulsation of the aorta, or of the other arteries in the body.

The patient died recently, and the diagnosis of carcinoma growing in the wall of the stomach was confirmed by autopsy.

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PAROXYSMAL VENTRICULAR TACHYCARDIA WITH RHYTHMIC ALTERNATION IN DIRECTION OF THE VENTRICULAR COMPLEXES IN THE ELECTROCARDIOGRAM

Report of Two Cases and Review of the Literature*

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INTRODUCTION

PAROXYSMAL ventricular tachycardia is a rare event and certain anomalies of this condition are still more rare. In the past thirteen years at the Massachusetts General Hospital there have been recorded electrocardiographically ten cases of paroxysmal ventricular tachycardia, as compared to sixty-six cases of auricular paroxysmal tachycardia, among 8,631 patients. One anomalous type of paroxysmal tachycardia of considerable interest, which has been found recently (within two years) at the Massachusetts General Hospital in three cases, consists of alternation in the shape of the abnormal ventricular complexes indicating varying distribution or origin of the abnormal beats. These three cases are the only ones of the group of ten seen at the Massachusetts General Hospital which showed this anomaly. In one case previously reported by Jones and White[12] the ventricular complexes are of two forms, only slightly divergent, appearing alternately during the paroxysm. In the two new cases here reported there is also alternation in form, but the difference is more marked. The ventricular complexes in the electrocardiograms successively alternate in direction and length of cycle during the paroxysm. This finding is not only of importance clinically because of its rarity and grave prognosis but also it is interesting from the standpoint of the underlying mechanism.

REVIEW OF THE LITERATURE

Paroxysmal ventricular tachycardia has been amply discussed in the literature; the criteria for its diagnosis electrocardiographically and clinically have been established[1, 2, 3, 4] and new cases gradually are being added to the rather small total number. The not infrequent relation of this condition to coronary sclerosis, coronary thrombosis (as in one case of this report), or focal myocarditis has been shown.[5, 9, 10, 11] Experimentally it has been demonstrated that ventricular ex-

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trasystoles, paroxysmal ventricular tachycardia, and ventricular fibrillation follow ligation of the coronary arteries.[5, 6, 7, 8]

It is known that the ventricular complexes of paroxysmal ventricular tachycardia may vary in time relation or length of cycle[4] and sufficiently in amplitude and shape to suggest origin from different ectopic foci.[12, 13, 14] Gallavardin has published tracings showing bizarre combinations of auricular arrhythmia and ventricular tachycardia.[15]

In 1911 Levy and Lewis,[16] working on cats under chloroform anesthesia with low chloroform tensions, showed that under these conditions injection of adrenalin chloride intravenously was followed by an increasing number of ventricular extrasystoles which were at times coupled. There then ensued a ventricular tachycardia showing regular continued alternation in direction of complexes, which changed into a series of very irregular abnormal beats and finally into ventricular fibrillation. The explanation given for the experimental curves was the origin of impulses from multiple ventricular foci, and the end-result of such a dominance of ectopic foci was ventricular fibrillation. In 1922 Schwensen[17] reported two cases under full doses of digitalis, the first showing coupling due to ventricular extrasystoles which were said to arise from multiple foci, and the second, a case of rheumatic mitral disease with auricular fibrillation, showing paroxysmal ventricular tachycardia with alternation in direction of complexes. No coronary thrombosis was found in either case at postmortem examination. Felberbaum,[20] in 1923, reported another case—a patient with a badly failing arteriosclerotic and hypertensive heart with auricular fibrillation who was given "heavy" doses of digitalis and who showed paroxysms of bidirectional, alternating, ventricular tachycardia. The patient died twenty-five days after the onset of the paroxysms. Reid,[21] in 1924, collected five cases in which paroxysmal ventricular tachycardia was caused, he felt, by overdigitalization, the condition having been confused in some cases with auricular flutter. In one case (Case 3 of his series) the electrocardiograms show the remarkable sequence of an alternating bidirectional ventricular tachycardia passing into ventricular fibrillation when death occurred. Luten,[23] in 1925, reviewed the subject and found two cases similar to those of Schwensen and Felberbaum and one in which alternation was not entirely rhythmic. In each case large amounts of digitalis had been given. Gallavardin,[25] in 1926, reported four cases, in at least three of which continued alternation in direction of complexes during ventricular tachycardia occurred. Gilchrist,[26] in 1926, reported five cases of paroxysmal ventricular tachycardia, in one of which for a few cycles at the beginning of one record a rhythmic alternation in direction of complexes was seen. Tobacco seemed a not unlikely

cause. Howard[27] reviewed the tachycardias due to digitalis in February, 1927, without adding any new cases of the bidirectional alternating type.

To summarize (Table 1).—There have been reported in the literature eight cases of continued alternation in direction of complexes in ventricular paroxysmal tachycardia. Six of the patients have died within from a few minutes to a few days of this finding, one lived twenty-five days after its onset, and one patient lived five months after it was first found. In connection with these cases, three instances of brief groups of alternating bidirectional ventricular complexes have been reported. Two of these patients died, and one was discharged well and could not be traced.

Some of the cases reported heretofore have been associated with large or even toxic doses of digitalis. In this connection reference should be made to the work of Gold and Otto[28, 29, 30, 31] who have shown clinically and experimentally that digitalis is not less potent or more dangerous in the presence of coronary thrombosis; that ventricular extrasystoles, spontaneous or induced by digitalis, do not indicate susceptibility to digitalis bigeminy, and finally that digitalis in full doses appears sometimes to abolish ventricular premature beats. The mode of action of the drug is a matter of conjecture; possibly it is due to the vagal stimulating effect. On the other hand atropine reduced or abolished coupled rhythm.[29]

Origin of impulses from separate ectopic foci, alternating right and left bundle-branch block, parasystole, re-entry, and circus movement have been proposed as the mechanism of alternating bidirectional paroxysmal ventricular tachycardia. These hypotheses will be considered below.

CASE REPORTS

CASE 1.—H. B. S.,* West Medical 287530, aged fifty years, native, married, male, superintendent of a paint factory, entered the Massachusetts General Hospital November 21, 1927 and died November 23, 1927.

Diagnosis: Arteriosclerotic and hypertensive heart disease. Angina pectoris. Cardiac asthma. Coronary occlusion.

Past History: There was an indefinite history of rheumatic pains in knees and shoulders. Typhoid fever twenty-five years ago. Some indigestion as a youth and again more recently. He suffered an attack of influenza in 1918. He also had gonorrhea twenty-five years ago. No history of lues.

Present Illness: He had played tennis until three years ago at which time he was obliged to cease playing because of gradually increasing shortness of breath and attacks of substernal oppression on exertion. At times he had had very slight swelling of the ankles. Seven months ago he was awakened by a severe substernal constricting pain radiating into both arms and bringing with it a feeling of suffocation and a sense of impending death. This attack, relieved by his local doctor, recurred once within the same twenty-four hours.

*The patient was sent to the hospital by Drs. A. L. Flanders and John L. Ames, of Boston, to whom we are grateful for information about him.

The patient was known to have had hypertension for several years and suffered at intervals from paroxysmal dyspnea, probably cardiac asthma.

On November 13, 1927, eight days before admission to the hospital, he complained of severe substernal and epigastric oppression "like inhaling strong ammonia," lasting two days, unrelieved by nitroglycerin, and requiring morphia. During this attack his blood pressure fell from 200 mm. mercury systolic and 100 mm. diastolic to 124 mm. systolic and 90 mm. diastolic. The patient was very sick both before and after entrance to the hospital, though up and around at first. The heart showed no abnormalities on routine examination two days after the severe attack. Gall-bladder disease was suspected and x-ray examination confirmed its presence.

Physical examination on admission to the hospital eight days after the onset of the present acute illness, showed an extremely ill, orthopneic, slightly cyanotic, large, obese man. His throat was slightly reddened, his chest was emphysematous,

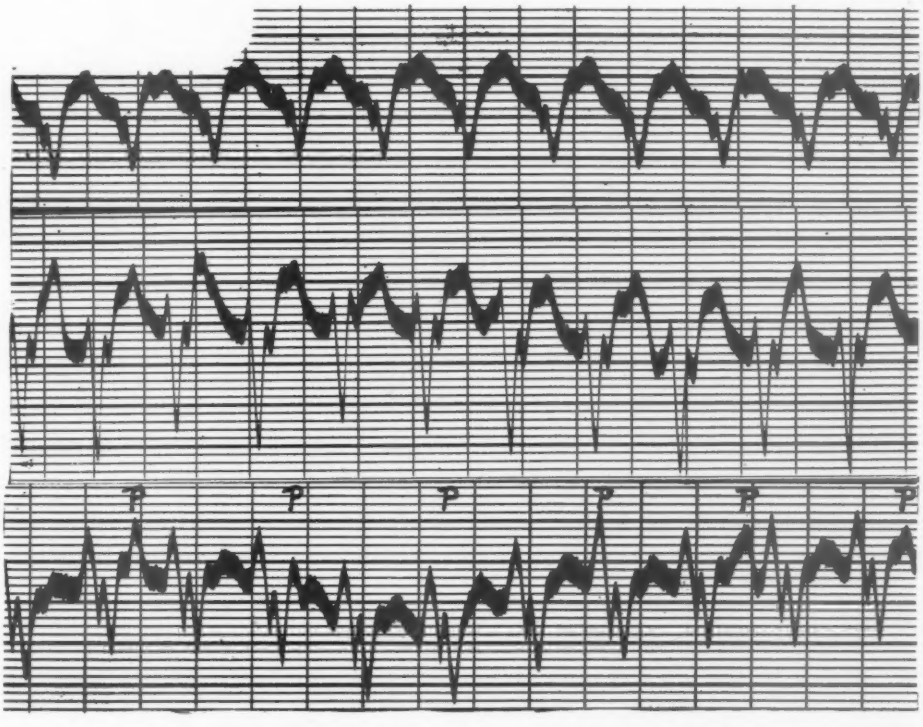

Fig. 1.—Case 1. Plate 13,892. Leads I, II, and III. Paroxysmal ventricular tachycardia with auricular rate 110, ventricular rate 193. Note P-waves superimposed on QRS and T-waves. Ventricular complexes are uniform in shape except when deformed by superimposed P-waves. Ventricular rhythm regular. (See Table II.) (Time interval equals 0.2 second, amplitude expressed by ordinates marking off 10^{-4} in all figures.)

and there were a few râles at the left base; the abdomen was negative. There was slight pitting edema of the feet. The heart was enlarged moderately to the left. The supracardiac dullness was slightly increased. The apex rate was over 190, with the sounds tic-tac in quality. No murmurs were heard. The radial pulse was weak and irregular at a rate of 80 to 100.

The first electrocardiogram was taken on the afternoon of the second day in the hospital (Fig. 1). The morning of the third day in the hospital the remaining electrocardiograms were taken (Figs. 2 and 3).

The patient was seen by us, as cardiac consultants, at noon of the third day in the hospital and a diagnosis of coronary thrombosis, angina pectoris, probable

cardiac asthma, and paroxysmal ventricular tachycardia was made. The points especially considered in addition to the history were temperature (101°), leucocytosis (25,000), and fall in blood pressure.

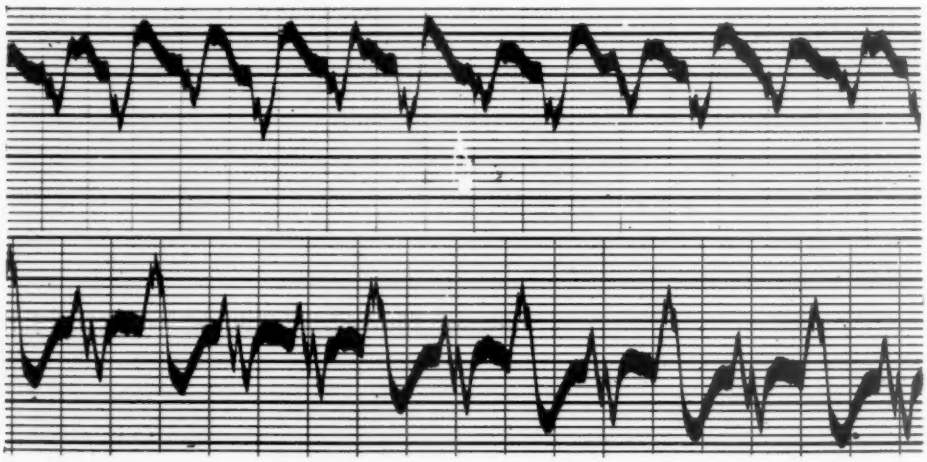

Fig. 2.—Case 1. Plate 13,906A. Leads I and III. Ventricular paroxysmal tachycardia with ventricular complexes of two different forms, both very abnormal and generally alternating in rhythm. In Lead III they are largely of opposite direction; in Lead I of similar direction but of different shape and amplitude. Note that the eleventh and twelfth complexes of Lead I and the fourth and fifth complexes of Lead III are similar, thus breaking at those points the exact alternation of the rhythm.

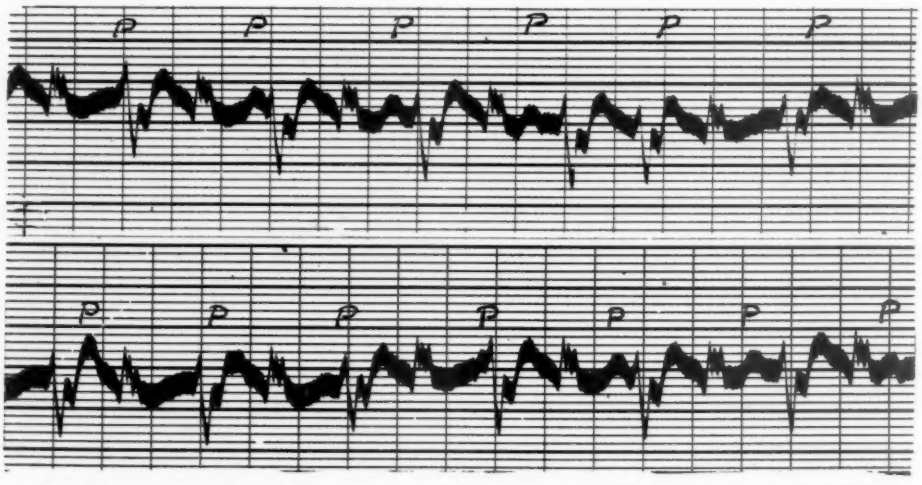

Fig. 3.—Case 1. Plate 13,906. Lead II. Same ventricular paroxysmal tachycardia as in Fig. 2. Alternate ventricular complexes are largely of opposite direction. P-waves superimposed on ventricular complexes. Auricular rate 110. Ventricular rate 210. Note that the eighth and ninth complexes of the first strip of Lead II are similar.

Laboratory Findings: The urine showed a slight trace of albumin, otherwise it was normal. The leucocyte count was 25,000. The Wassermann reaction was negative. The icteric index of the blood was 5.

Digitalis Treatment: For three weeks before entrance the patient received 1-2 minims of the tincture of digitalis per day and for the last three days before entrance one ampule intramuscularly, of a digitalis preparation (corresponding to gr. iss of the digitalis leaves) per day. In the hospital, he received digitalis leaves

gr. ivss the first day, gr. xviii the second day and gr. iii the third day (a total of 25½ grains or 1.7 grams). This was not an overdose. The treatment otherwise was absolute rest, with morphia and nitroglycerin for pain.

The patient died suddenly on the afternoon of the third day, the rapidity of his apex rate not having once abated while in the hospital.

Necropsy 5252*: Positive findings. Firm fibrous adhesions of omentum and transverse colon to gall bladder were found. There were 300 c.c. of clear fluid in each pleural cavity. The lungs showed marked congestion and edema.

The pericardium contained 30 c.c. of turbid semipurulent fluid. There was a slight fibrinous exudate on the posterior surface of the left ventricle.

The *heart* (Fig. 4) weighed 545 grams. It was considerably enlarged; the cavities were dilated. The apex of the left ventricle felt distinctly soft and showed several patches of discoloration, some bright red, others white. On section, necrosis of the lower half of the posterior and lateral walls of the left ventricle was found.

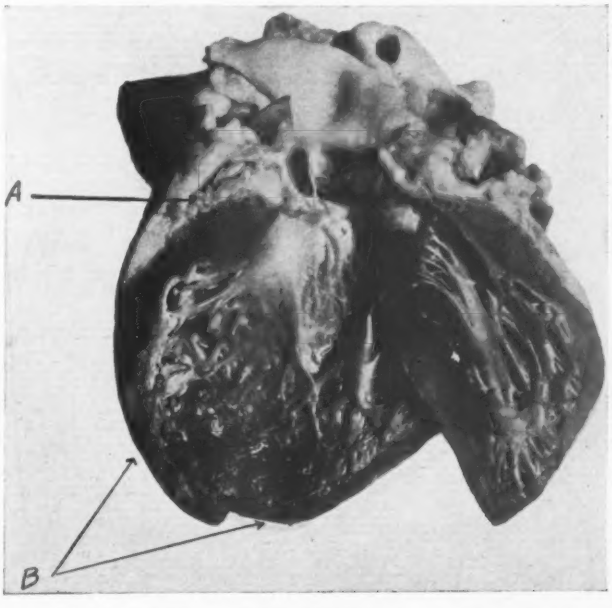

Fig. 4.—Heart of Case 1. Showing marked thinning and necrosis in the large area of infarct of the left ventricle and large mural thrombus. *A*, exposed and cut left descending coronary artery. Arrow indicates point at which arterial thrombus began. *B*, extensive necrosis and large mural thrombus.

The average thickness of the ventricular wall above the area of necrosis was 12 mm. and in the area of necrosis 8 mm. The thickness of the right ventricle was 5 mm. All the valves were negative except for slight calcification and a few atheromatous plaques. There was marked atheroma of all branches of both coronaries; the right, however, was not diminished in diameter. The circumflex branch of the left coronary artery showed several areas of calcification which encroached considerably on the lumen. The descending branch of the left coronary was markedly narrow, and at a point 2.5 cm. from the orifice there was a fresh pinkish thrombus. Below this thrombus the lumen was almost completely obliterated for a distance of 1.5 cm.

The aorta was negative aside from moderate sclerosis of the arch and descending portions.

*By Dr. Tracy Mallory.

The kidney capsules stripped with difficulty, leaving surfaces that showed numerous deep pitted scars and also a small amount of fine granulation.

CASE 2.—A. J. H., East Medical 284540. A white, male, married, Syrian laborer, aged forty-five years.

Diagnosis: Cardiac enlargement and weakness of unknown cause. Auricular fibrillation, ventricular paroxysmal tachycardia, congestive failure.

History: He suffered his first attack of heart failure and entered the Massachusetts General Hospital six and one-half years before his second admission. At that

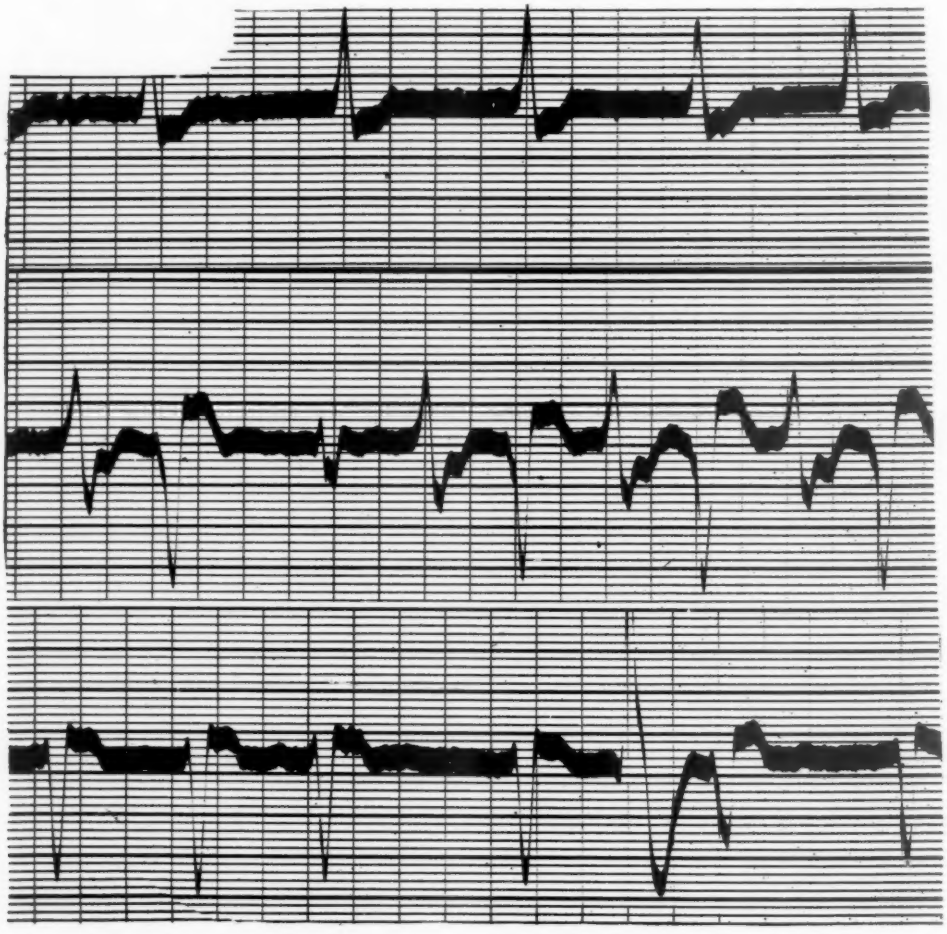

Fig. 5.—Case 2. Plate 13,281. Leads I, II, and III. Auricular fibrillation and periods of trigeminy. In Lead II occurs the onset of a paroxysm of ventricular tachycardia with ventricular complexes alternating in direction and of exactly the same shape and time interval as in the case of the trigeminal ectopic contractions. The heart rate during the fibrillation 80-100 and during the paroxysm of tachycardia 152.

time a diagnosis of chronic myocardial insufficiency, left hemiplegia and questionable coronary occlusion with mural thrombus and cerebral embolus was made. On physical examination the heart was large. There was a systolic murmur at the apex. The blood pressure was 130 mm. mercury systolic and 70 mm. diastolic. His electrocardiogram at that time showed, in addition to ventricular extrasystoles, auricular fibrillation, left axis deviation, and a ventricular rate of 115. He had a slight fever. The Wassermann reaction was negative.

The patient entered the hospital the second time on June 29, 1927 and died July 13, 1927. In the interval of six and one-half years he had enjoyed fair health until two weeks before admission, when his abdomen began to swell, his heart became "jumpy," and he suffered from pain over the precordium. One week before admission he became very edematous and complained of cough. There was no history indicating coronary occlusion. There was no rheumatic history but he had yearly attacks of severe sore throat.

Physical examination showed a well-developed and nourished cyanotic man with Cheyne-Stokes respiration. The heart was huge. There were apical and basal systolic murmurs and a gallop rhythm. The pulse was absolutely irregular. The blood pressure was 135 mm. mercury systolic and 100 mm. diastolic. The liver was large and tender. Sacral edema and edema of the lower extremities was present. Lungs showed moisture at the bases.

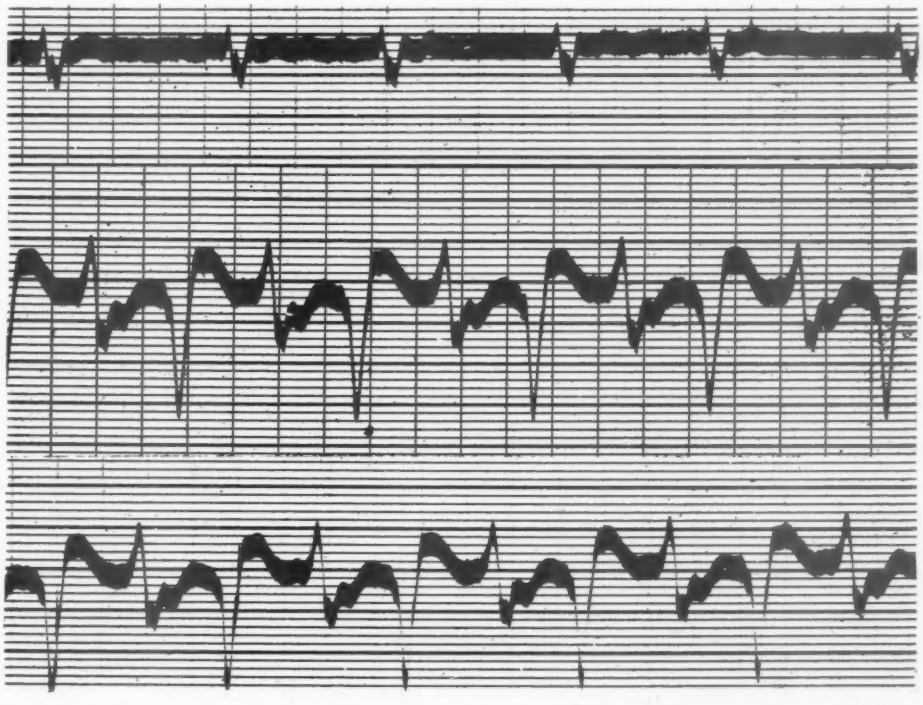

Fig. 6.—Case 2. Plate 13,282A. All Lead II. Showing in first strip auricular fibrillation at a rate of 80. Second and third strips show ventricular paroxysmal tachycardia as described in Fig. 5.

In the first two days in the hospital he received thirty grains of digitalis and one and one-half grains daily thereafter for ten days. He showed a fall in pulse rate from 105 to 70. A deficit of from five to ten beats persisted. There was no evidence of digitalis intoxication.

The patient did very poorly, developed a right facial and right arm paralysis, and finally bronchopneumonia, and died. No autopsy was obtained.

The electrocardiograms (Figs. 5 and 6) were taken on the third day in the hospital.

DISCUSSION

The alternation in direction of the complexes in paroxysmal ventricular tachycardia so far encountered has borne an extremely bad prognosis; it has been associated with serious heart disease and con-

gestive or anginal failure. Some of the cases thus far reported apparently have received very large or even toxic doses of digitalis. Our cases were fully digitalized without toxic effects having been noted. We believe that digitalis was indicated in our patients. The work of Gold and Otto[28, 29, 30, 31] referred to above has shown experimentally that it is not a dangerous drug in extrasystolic irregularities. Nevertheless it appears to be true that in some cases, namely Schwensen's second case, Reid's case showing alternation in direction, and Luten's second and third cases, the patients received an excessive amount of digitalis. In the other cases reported, the amounts of digitalis given are not clearly indicated.

In the total series of thirteen cases (Table I), ten of them including our own two new cases, showing continued alternation of direction, arteriosclerotic and hypertensive heart disease was present four times, chronic nephritis twice, coronary thrombosis once, coronary sclerosis without hypertension once, luetic heart disease twice, and rheumatic heart disease once; cardiac asthma occurred once, anginal failure twice, congestive failure eleven times; some cases having more than one of these conditions present. There was little or no congestive failure in the one case discharged well. The age distribution was from forty-one to eighty years, averaging fifty-nine and six-tenths years. A badly nourished myocardium appears to be a factor; overdosage with digitalis may also be a precipitating cause through the action of the drug on the heart muscle.[21, 22]

The *mechanism* underlying this disorder is a subject for theoretical consideration and various possibilities will be considered.

1. The origin of the impulse of the tachycardia from a single focus, or, as will be discussed below, from a circus movement or continued re-entrant waves, may be above the bifurcation of the bundle of His and the alternating direction be the result of *alternating right and left bundle-branch block*, due to refractory right and left bundle-branches, respectively. This is, however, improbable, as the beats differ in time of appearance. Changing and variable bundle-branch block has been observed[24] but with regular rhythm unless fibrillation was present.

2. Again, if we suppose the impulse to arise from a single focus above the bifurcation of the His bundle, it is conceivable that the *Purkinje fibers* of the ventricles themselves may be so *refractory* in such masses preponderantly in alternate ventricles with successive beats that the refractory states of the two ventricles alternate in their effects. The side of the heart with most fibers conducting impresses the cardiogram of that side in the tracing, the aberrant levogram and dextrogram thus alternating. Here too, however, the rhythm should be a regular one and not alternating in time as we find it in our cases.

Also, if anything, this mechanism is less likely than that cited in the first paragraph, just as arborization block is less likely than partial bundle-branch block.

3. Though aberration may result from rapidity of a tachycardia, the alternate types of aberration discussed here suggest that the spread of the excitation is in opposite directions in the muscle relative to the position of Lead II. Origin of the ventricular tachycardia from *two separate foci* would thus seem not unlikely. When the beats occur inaccurately related to each other, this is very probable, according to the parasystolic theory of Kaufman and Rothberger.[33] This theory supposes that extrasystoles result from the interplay of two separate rhythms, as is obvious, for instance, in the patient reported by one of us (P. D. W.[36]), showing idioventricular rhythm occasionally interrupted by an impulse coming down from the auricles when the auricular stimulus falls at a suitable point in diastole. Certain cases of extrasystole with inaccurate coupling have been shown to have a simple mathematical relation between interextrasystolic periods. Cases showing accurate coupling have been explained on the basis of parasystole by interference between the two rhythms resulting in allorhythmia with uniform bigeminy. Sherf[37] conceives that there is not only a protective blocking of the ectopic-rhythm-producing center so that its ectopic rhythm will not be interfered with, but that there is a block preventing impulses from going out and that the effect of the parasystolic rhythm on the normal rhythm waxes and wanes. By the use of these ideas almost any rhythm can be explained.

In our two cases and in the other similar cases referred to in the literature parasystole may be the mechanism. With such extensive myocardial damage as was found in Case 1 certainly more than one focus may have been formed. This seems, however, unlikely because of the exact equality of the rates of stimulus production in the two supposed foci. Circus movements might also exist in such areas throwing off impulses with variable responses on the part of the ventricles. Sherf[37] indeed believes there may be a continuously contracting, refractory "fence" around areas producing ectopic rhythms.

4. *Re-entry* is a possible explanation of extrasystolic phenomena and has been fully discussed by Lewis.[22] An example of reciprocal beating in a case observed by one of us (P. D. W.[32]), where the R-P interval in A-V dissociation widened to a certain point at which the P-wave gave rise to a ventricular complex, illustrates the likelihood of the mechanism of re-entry. Many instances are known of successive responses to a single stimulus and of continued multiple responses after a series of rhythmic extrasystoles have been forced. These phenomena are likely to occur when the stimuli fall early in diastole just beyond the usual refractory period but while the muscle is in a partially

TABLE I

A SUMMARY OF THE FINDINGS IN THE PREVIOUSLY REPORTED CASES TOGETHER WITH OUR OWN. THE COLUMNS MARKED *Extrasystole* AND *Coupling* REFER ONLY TO THE FINDINGS IN THE PUBLISHED CURVES. UNDER THE HEADING *Cycle Length* ARE TABULATED THE ROUGH CALIPER MEASUREMENTS FROM THE PUBLISHED RECORDS. SOME OF THE CASES, AS INDICATED, APPEARED TO VARY CONSIDERABLY IN CYCLE LENGTH, IN OTHERS CYCLE LENGTH ALTERED AS DID DIRECTION OF THE VENTRICULAR COMPLEXES. SOME SHOWED BOTH ALTERNATION IN LENGTH AND REGULAR EQUAL LENGTHS, WHILE ONE CASE SHOWED BOTH OF THESE AND IN OTHER PLACES CONSTANT VARIATION. IN THE LAST COLUMN BUT ONE IS NOTED WHETHER THE STRING WAS IN CONTINUOUS MOTION INDICATING PERHAPS CONTINUAL MOVEMENT OF THE EXCITATION WAVE OR WHETHER THERE WERE ISO-ELECTRIC PERIODS.

AUTHOR—REF.	AGE	DIAGNOSIS	DIGITALIS	AURICULAR FIBRILLATION	BIDIRECTIONAL ALTERNATION		EXTRA-SYSTOLES		COUPLING		CYCLE LENGTH			BASE LINE		LIFE AFTER ONSET OF ALT. DIRECT. TACHYCARDIA
					BRIEF	CONT'D	SINGLE FOCUS	MULTIPLE FOCI	ACCURATE	INACCURATE	EQUAL	ALT. EQUAL	VARIOUS	CONT'D MOTION	ISO-ELECT. PERIODS	
Schwensen [17]	61	Rheum. Mitral Dis. Cong. Failure	4.85 gm. in 13 days	+		+	0	0	0	0		+		+		5 days
Felberbaum [20]	54	Art. Hyper. Heart Dis. Cong. Failure	"Heavy" doses	+		+		+	+		+		+	+	+	25 days
Reid [21]	80	Art. Hyper. Heart Dis. Cong. Failure	? previous amount. 23 c.c. tr. in 2 days wt. 135 lb.	+		+	0	0	0	0	+				+	a few seconds
Luten [23]																
Case 1	64	Chr. Myocarditis Cong. Failure	12 c.c. tr. in 24 hr.	+		+	0	0	0	0	+	+	+	+	+	24 hours
Case 2	44	Thyro. Cardiac. Coronary Sclerosis Cong. Failure	24 c.c. tr. in 8 hr.	+		+		+	+	+	+			+		24 hours
Case 3	71	Art. Hyper. Heart Dis. Chr. Nephr. Cong. Failure	19 c.c. tr. on 7th day	0	+		+		0	+		+			+	2 hours

TABLE I—CONT'D

Gallavardin	25 1	77	Anginal Failure. ? Uremia	*	+		+	+	+	0	0				+		5 mo.
	2	56	Luetic Ht. Disease Aortitis Cong. Failure	"Full digitalis"	0		+		+	+		+				+	3 days
	3	70	"Nephroaortic Syphilis" Aortic insufficiency Cong. Failure	*	0		+	0	0	0	0	+				+	"a few days"
	4	62	? Primary Hypert. ? Myocard. infaret. ? Luetic Ht. Disease & Aortitis Cong. Failure	*	0	+		0	0	0	0		+†		No cont'd paroxysm.		7 days
Gilchrist	26	41	Parox. vent. tachycardia	No digitalis	0	+		0	0	0	0	+			+		Disch. well
Present Report	1	50	Art. & Hyper. Ht. Disease Angina Pectoris. Card. Asthma. Coronary Occlusion Cong. Failure	1.7 gram in 3 days	0		+	0	0	0	0		+		+		6 hr.
	2	45	Cardiac En'largement and Weakness of Unknown Cause. Cong. Failure. Vascular Crisis	2 grams in 2 days and 0.1 gm. daily for 10 days	+		+	+		+			+		+		13 days

*Records are brief summaries and do not give clearly the relation of digitalis to the tachycardia.
†Corresponding parts of the different couples were equal to each other.

refractory phase, due to malnutrition, disease, or vagal stimulation. The nature of such multiple responses in the auricle is known to be a state similar to flutter or fibrillation. As Lewis points out, the theory of re-entry and of circus movement as an explanation of extrasystoles and paroxysmal tachycardia is more applicable to the ventricle than to the auricle for in the ventricle conduction is slower, the refractory period is longer, and the chamber is larger. The wave has the time and the space in which to circulate. Moreover, the complexes are often contiguous or at least the string is seen to be in continuous movement, as in our two cases. Finally, the relation between extrasystoles, paroxysmal tachycardia and ventricular fibrillation, as noted above, is very close.[5, 9, 10, 11, 21]

Circus movement in the ventricle has not been proved, but Lewis[7] has observed that in the fibrillating ventricle of the dog there appears to be an underlying movement aside from the fibrillary twitchings. The two cases here reported, and other cases showing equal or regular cycle lengths, especially those with no iso-electric periods in the electrocardiograms, may represent a continuous activity of the ventricular muscle, a ventricular flutter. The occasion of such a condition, if it exists, by analogy with circus movement in the auricle, probably depends upon a state of partial refractoriness. The very fact that irregularity of rate is common and well recognized in paroxysmal tachycardia of ventricular origin favors the idea that a circus movement is responsible, the path of the circus varying somewhat from time to time, as it does to a lesser degree in auricular flutter, and thus accounting for the variation in time and also in shape of the abnormal ventricular complexes. Fischer[38] has recently called attention again to this variation in time and shape which he ascribes to the activity of two separate ventricular foci. His explanation, however, appears less likely than that given above; we have already discussed this point.

The oppositely directed complexes of our cases indicate the tendency to alternating left ventricular and right ventricular direction of the hypothetical circus movement, a figure-of-eight arrangement.* A rough measurement of the outside apex-base circumference of the heart in Case 1 was 294 mm. At the rate of 400 mm. per second an impulse traveling in the muscle alone would complete such a circuit in 0.735 second; this interval of time, even if much shortened by passage along some of the Purkinje network, might still be as great as the 0.310 second interval found in Fig. 1. There could thus be time enough for such a circus.

An important finding in our two cases is the result of accurate measurement of the R-R intervals (by the Lucas Comparator). We find

*It is of special interest that in Case 2 the tachycardia consists of a constant repetition of the two oppositely directed ectopic beats occurring as part of a trigeminal rythm during the intervals between the paroxysms of tachycardia; thus a single figure-of-eight circus wave may explain the trigeminy. See Fig. 5.

TABLE II

THE RESULT OF MEASUREMENT OF TIME INTERVALS* WITH THE LUCAS COMPARATOR OF ELECTROCARDIOGRAMS OBTAINED FROM THE TWO CASES HERE REPORTED. (SEE TEXT FOR DISCUSSION OF THIS TABLE.)

CASE 1. FIG. 1. LEAD III, Q-R-S 2-12 INCLUSIVE (NO. 12 NOT SHOWN IN FIGURE)		CASE 1. FROM LEAD II, UNPUBLISHED RECORDS		CASE 1. FIG. 2. LEAD III, Q-R-S 1-11 INCLUSIVE		CASE 2. FIG. 5. LEAD II, Q-R-S 1-14 INCLUSIVE (NOS. 10-14 NOT SHOWN IN FIGURE)	
RELATION OF COMPLEXES	R-R INTERVAL	RELATION OF COMPLEXES	R-R INTERVAL	RELATION OF COMPLEXES	R-R INTERVAL	RELATION OF COMPLEXES	R-R INTERVAL
– –	0.3156	– +	0.2786	+ –	0.3054	+ –	0.4150
– –	0.3078	+ –	0.3120	– +	0.2906	– N	0.6966
– –	0.3090	– –	0.3136	+ –	0.3112	N +	0.4342
– –	0.3110	– +	0.2664	– –	0.2982	+ –	0.4116
– –	0.3098	+ –	0.3202	– +	0.2880	– +	0.4226
– –	0.2948	– +	0.2838	+ –	0.2966	+ –	0.3800
– –	0.3190	+ –	0.3108	– +	0.2998	– +	0.4080
– –	0.3114	– +	0.2826	+ –	0.3042	+ –	0.3858
– –	0.3074	+ –	0.3108	– +	0.2876	– +	0.4072
– –	0.3992	– +	0.2772	+ –	0.3066	+ –	0.3718
Average	0.3095	– + Average	0.2777			– +	0.4128
Rate	193	Rate	216	– + Average	0.2915	+ –	0.3784
		+ – Average	0.3144	Rate	205	– +	0.3982
		Rate	190	+ – Average	0.3058		
				Rate	196	– + Average	0.4099
						Rate	146
		Combined		Combined		+ – Average	0.3855
		Average	0.2966	Average	0.2988	Rate	152
		Rate	210	Rate	201		
						Combined	
						Average	0.4250
						Rate	141

*In seconds.
N Natural complex; N– Interval from invert to natural complex;
N Interval from natural to upright complex.
+ – Interval between an upright complex and an inverted complex.
– + Interval between an inverted and an upright complex.
– – Interval between two successive inverted complexes.

in the electrocardiogram of Case 2 (Fig. 5, and Table II) that with the onset of the paroxysm there is not only alternation in direction of complexes but also there is alternation in length of cycles, the interval between an inverted and an upright complex being definitely longer than the interval following an upright complex. How closely each alternate interval approximates other corresponding intervals is more apparent when one observes that the paroxysm at the onset is increasing in rate for the first few cycles. The significance of this is increased by the observations on the different rates in Case 1. In Table II the R-R intervals of Lead III (Fig. 1), all inverted, are tabulated. The average length is 0.3095 second, the rate 193. The next morning alternating bidirectional ventricular tachycardia had set in. The electrocardiograms shown in Figs. 2 and 3 were obtained. Reference to Table II and to Fig. 2 shows that again we have alternation in R-R intervals, the interval following an inverted complex being in this instance definitely shorter than the interval following an upright complex. Finally with the onset of the alternating tachycardia the rate rises from 193 to 210 per minute. It seems quite possible that these two fairly constant, distinct time intervals with constant and distinct forms of ventricular complexes represent two circus movements, largely in opposite directions. On this hypothesis the new interrupting circuit would be represented in Case 1 by the upright complexes which appear in Leads II and III of Figs. 2 and 3. These show a tendency to right axis deviation, and on the hypothesis would result from a more rapidly beginning circuit directed towards the right, possibly into the right ventricle. The circuit represented by the inverted complexes would correspondingly be directed to the left, probably in the left ventricle. The hypothesis would suppose, then, the interplay of two ventricular circus movements, the tendency to orderly successive right and left cardiograms being caused by a double, or figure-of-eight, circuit. It may be expressed as follows: The circus movement takes whatever path it can find. Presently it finds the path it had been following primarily in the left ventricle refractory, and it pursues a course now primarily towards the right. We might suppose the impulse wave to leave the primary left ventricular circuit at some time before completing the entire circuit, thus accounting for the shorter interval following the inverted complexes. The interrupting or secondary circuit on its return finds the primary path open and the left ventricular circus movement then resumes its way until interrupted again by the new circus in the other direction. On this hypothesis there would be a single figure-of-eight circus through the ventricles, each causing two complexes resembling successively dextrocardiograms and levocardiograms.

It is apparent that any plane drawing, in an attempt to represent visually the possible path of this hypothetical ventricular circus, is a

very rough approximation of the complicated and tortuous course which such a circus movement must pursue. Indeed, reference to the two alternating complexes as "right" and "left" must also be considered as somewhat approximate. It may be stated more justly that, considering the heart as a whole, the inverted complexes represent excitation from the apex toward the base and that the upright complexes represent excitation from the base toward the apex. We have no evidence to indicate the exact path of the excitation wave. It may be through the chambers in any one of several planes and directions; it may, and quite likely would traverse the septum.

It is interesting that though alternation in form occurs, there is no alternation in direction in Lead I. The answer lies in the relation

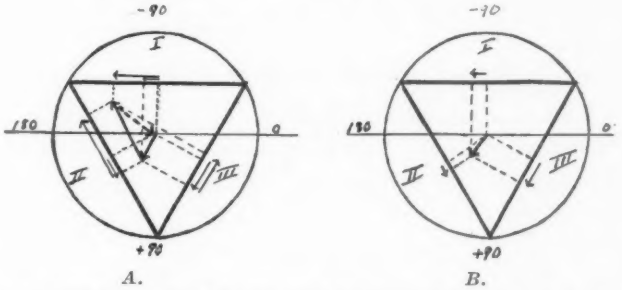

Fig. 7.—The approximate angles for the electrocardiograms of Case 1, shown in Figs. 1 and 2 are plotted in relation to Einthoven's triangle. Two phases are shown for Fig. 1, indicating in a general way the direction of the circus. The diagrams demonstrate the reason why both types of complex are inverted in Lead I.

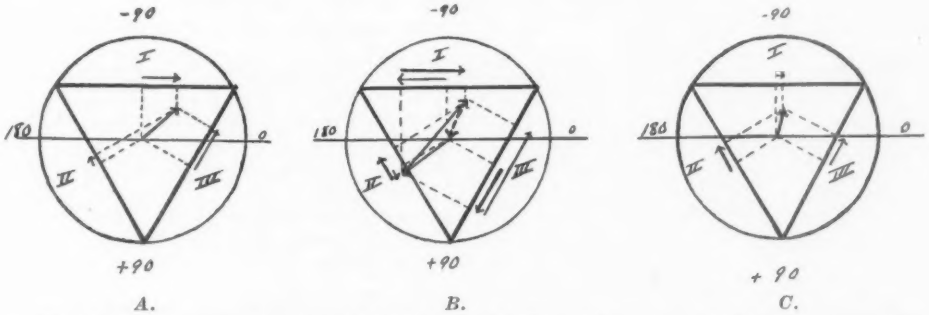

Fig. 8.—The approximate angles for the electrocardiograms of Case 2, shown in Figs. 6, 7, and 8, are plotted in relation to Einthoven's triangle, the successive complexes of the trigeminal groups of Fig. 7 taken as representative complexes. The first, second, and third complexes are shown in *A*, *B*, and *C* respectively.

of the plane of the probable circus movements to Lead I. We have plotted the measurements of the chief angles of the axis deviation for Figs. 1, 2 and 3 and they are shown in Fig. 7, *A* the first tachycardia and *B* the additional alternate upright complexes. It is clear from these diagrams why the complexes in Lead I are inverted. The plane of the circus, if such exists, indicated in Fig. 7-*A* is probably more frontal than that of the second circus. Fig. 8 shows the resultant axis deviations of the alternate ventricular complexes of Case 2.

Whether quinidine sulphate might act favorably or unfavorably on such a mechanism as this producing the bidirectional ventricular

paroxysmal tachycardia, we do not know. Because of the possibility of unfavorable toxic effects this drug, so far as we know, has not been used in such cases.

SUMMARY

1. Two cases of alternating bidirectional paroxysmal ventricular tachycardia are reported, with a review of the literature. The new cases reported alternate in cycle length as well as in direction.

2. Etiologically alternating bidirectional paroxysmal ventricular tachycardia seems to be associated with severe heart disease and congestive or anginal failure, and in some cases with excessive administration of digitalis.

3. Clinically these cases are of especial interest because of their bad prognosis.

4. We have considered several possible mechanisms for this unusual tachycardia and on the basis of careful measurement of the electrocardiographic plates have ventured to suggest the possibility of a double ventricular circus movement as a likely mechanism.

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THE ELECTROCARDIOGRAPHIC CHANGES IN PNEUMOTHORAX IN WHICH THE HEART HAS BEEN ROTATED

The Similarity of Some of These Changes to Those Indicating Myocardial Involvement*

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MERE displacement of the heart as a whole to the right or left produces little change in the electrocardiogram. Several observers[1, 2, 3, 4] have noticed this in hydrothorax. Nevertheless it is well known that the position of the heart does influence the electrocardiogram. This was first shown by Waler.[5] Einthoven, Fahr, and de Waart[6] analyzed the influence of respiration upon the electrocardiogram and suggested that rotation, rather than lateral movement, was the cause of the inversion of the QRS group in the third lead of the electrocardiogram in individuals with hypertrophy of the left ventricle. The importance of rotation around anteroposterior and longitudinal axes of the heart has been noted by Boden and Neukirch,[7] Herrmann and Wilson,[8] Hoffmann,[1] Waller,[9] Groedel[10] and others.

The changes in the electrocardiogram caused by moving a patient from the supine to the left lateral and then to the right lateral positions have been utilized electrocardiographically to indicate the presence or absence of an adherent pericardium.[11] Recently Meek and Wilson[4] have shown that in dogs rotation of the heart on an anteroposterior axis or around the longitudinal axis produces the usual electrocardiographic signs for right and left axis deviation (ventricular preponderance) of the heart.

Some confusion prevails concerning the effect on the electrocardiogram of cardiac displacement caused by air in the thoracic cavity. Lewis[2] and other workers[3] state that this condition produces very little or no change in the electrical tracing of the heart and this view seems to be held very widely. Yet Egan,[12] more than thirteen years ago, showed that in four cases of artificially produced right pneumothorax the amplitude of the S-wave in Lead I and of the R-wave in Lead III was increased. Although no electrocardiographic or x-ray records were published, these were taken in every case. The x-ray films were reported to have shown displacement of the heart to the left, and one diagrammatic sketch summarizes the changes in the electrocardiograms.

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This matter was called to our attention at the New York Hospital by an electrocardiogram taken on a nineteen-year-old boy which showed a simple tachycardia, with a rate of about 110 per minute, and a right axis deviation of the QRS group (Fig. 1). The S-T segment in the first lead was slightly above the iso-electric level and in the second and third leads it was below—changes suggesting myocardial damage of the type usually produced by a fairly recent coronary closure. There was no reason to suspect myocardial degeneration in this patient, particularly in view of his youth. An explanation for the electrocardiographic changes had to be sought and so his case was investigated in some detail.

CASE HISTORY

CASE 1 (271,208).—A nineteen-year-old English lad was admitted to the medical service January 28, 1927 with a signal symptom of pain in the chest. For one month there had been cough, progressive weakness, and anorexia. One week prior to

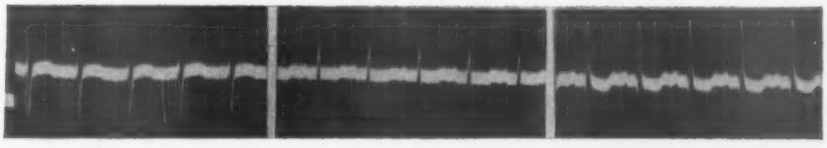

Fig. 1.—Case 1. Electrocardiogram showing simple tachycardia, rate of about 110 per min., a right axis deviation of the QRS group. Abnormalities of the S-T segment present. (Reduced one half.)

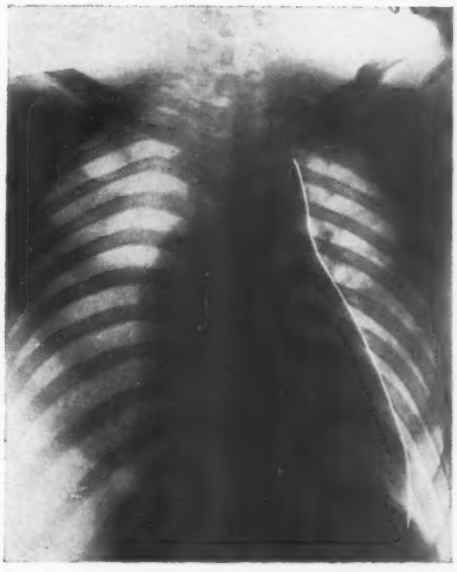

Fig. 2.—Case 1. X-ray picture showing complete right pneumothorax with displacement of mediastinum and heart to left.

admission he had developed a sharp pain just to the right of the sternum. The past history, family history, and habits were negative.

Physical examination revealed a poorly nourished, acutely ill, cyanotic young adult, lying on his right side, breathing rapidly. The entire right side was hyper-

resonant and the breathing was amphoric. This side was more prominent than the left and moved less on respiration. The examination of the heart showed no recognizable abnormality. The apex beat was forceful, just within the nipple line; there was no increase in dullness to the left. The pulses were equal, strong, full, and regular. The liver edge was palpable 2 cm. below the costal margin.

An x-ray film taken January 29, 1927, on the day after admission, showed a complete right pneumothorax (Fig. 2). The trachea, bronchi, and heart were displaced to the left. No definite diagnosis of tuberculosis was made. The electrocardiogram taken January 31, 1927, as already mentioned, indicated simple tachycardia, rate 110 per minute, a marked right axis deviation of the QRS group and abnormalities of the R-T or S-T segment in that, in the first lead this interval was slightly above the iso-electric level, and in the second and third leads, below (Fig. 1). The amplitude of the QRS group in the third lead was large. No digitalis

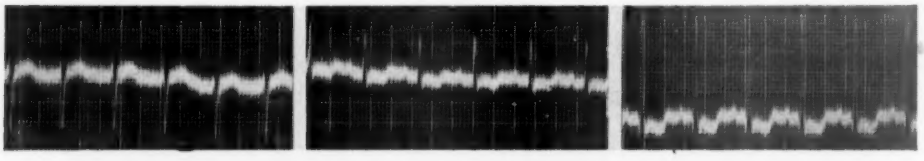

Fig. 3.—Case 1. Patient in supine position.

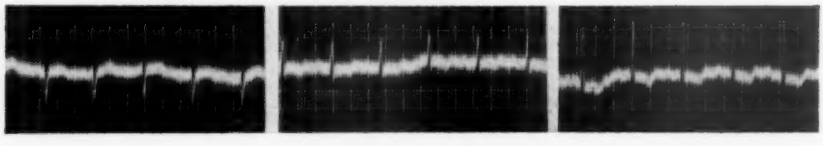

Fig. 4.—Case 1. Patient in left lateral position.

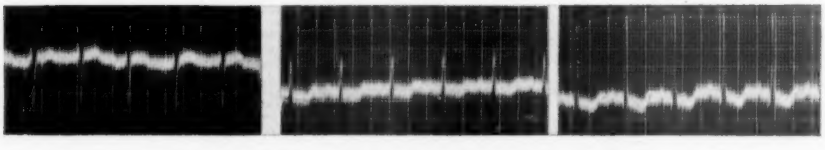

Fig. 5.—Case 1. Patient in right lateral position.

had as yet been given and the patient was now placed on this drug. More x-ray plates and electrocardiograms were taken. The P-wave in the electrocardiogram became more marked. Electrocardiographic tracings were taken February 15, 1927, in the supine and the two lateral positions (Figs. 3, 4 and 5). Only in the left lateral position was there a variation from the supine—a diminution in voltage. These facts are cited to show, at least circumstantially, that rotation of the heart had taken place. Presumably the organ was rotated to the right by the pneumothorax and hence, with the patient on that side, no further change was produced, whereas when placed on his left side, the heart was rotated back to the left. (The rotation to the right may have been either around a longitudinal axis of the heart or around an anteroposterior one.)

Later the patient developed a pyopneumothorax from which he died on March 25, 1927. The autopsy revealed, in addition to the right pyopneumothorax, a bilateral suppurative bronchopneumonia, a bilateral pulmonary tuberculosis, and a left fibrinous pleurisy. The pericardial cavity was normal; the heart weighed 275 gm. and was normal in size and shape. No focal lesions were found; the endocardium and valves displayed no abnormality. This normal condition of the endocardium,

myocardium, and pericardium is emphasized in order to establish the point that the marked electrocardiographic changes were produced by the *rotational effects of the pneumothorax* and not by any change in the heart muscle itself.

Two weeks later a patient was admitted who received artificial pneumothorax treatment. Electrocardiograms were taken before and after each injection of air.

CASE HISTORY

CASE 2 (272,409).—The patient was a Porto Rican man, twenty-nine years of age, a factory hand. He was admitted April 7, 1927 with chronic pulmonary tuberculosis and was discharged improved on August 14, 1927. To be brief, a bed-

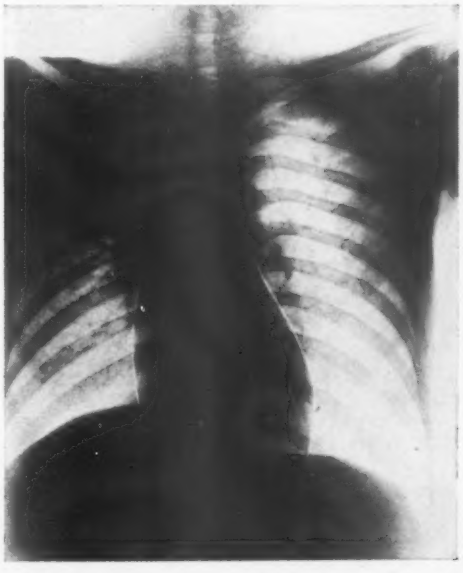

Fig. 6.—Case 2. Roentgenogram of chest before pneumothorax treatment showing mottled increase in density of right upper lobe.

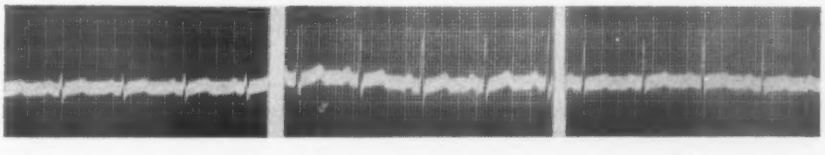

Fig. 7.—Case 2. Normal electrocardiogram, before pneumothrax treatment.

side x-ray film taken April 8, the day after admission, showed a mottled increase in density of the right upper lobe (Fig. 6). An electrocardiogram taken April 11 showed a normal record (Fig. 7). On April 14 an artificial pneumothorax was performed; 1000 c.c. of nitrogen being injected into the right thoracic cavity. The x-ray film showed that the entire right lung was fairly well collapsed; the upper lobe occupied the median two-thirds of the chest and was adherent to the chest wall; the lower lobe was completely collapsed (Fig. 8). A slight decrease in the voltage of the QRS group was immediately noted, and this was more marked four days later when the QRS group was still smaller and the T-waves had decreased in amplitude and had become flat in the third lead (Fig. 9). Slowly the electro-

cardiogram returned again to its original form, but further injections of air produced similar electrocardiographic changes.

The opportunity of following this patient presented itself October 24, 1927, when he was readmitted to the New York Hospital. The x-ray film showed a fibrotic change in the right lung with the trachea and mediastinum pulled over to the right (Fig. 10). No pneumothorax was present and the electrocardiogram was normal (Fig. 11).

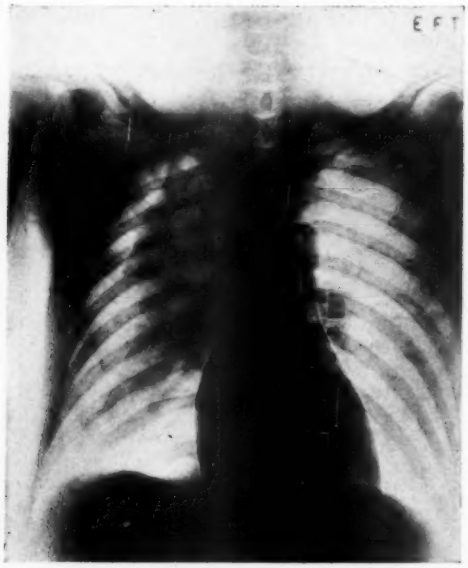

Fig. 8.—Case 2. Roentgenogram showing collapse of right lung due to injection of 1000 c.c. of nitrogen. Lower lobe quite collapsed.

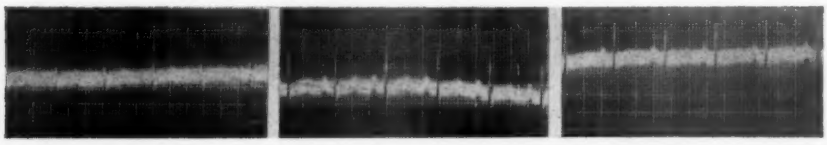

Fig. 9.—Case 2. Electrocardiogram taken after pneumothorax was induced. T-waves smaller. Voltage of QRS still lower. (Compare with Fig. 7.)

In this case, with no observable abnormality in the heart, definite electrocardiographic changes were noted whenever a pneumothorax was produced. These changes could be produced at will.

It is not necessary to discuss in detail the course of five other patients. One patient (Case 3) died within twenty-four hours of admission and showed at autopsy a spontaneous right pneumothorax. There was no opportunity for an x-ray film, but the electrocardiogram (Fig. 12) showed a tachycardia, low voltage of the QRS group and T-waves, and slight right axis deviation of the QRS group. These findings were present in spite of the fact that autopsy revealed no change in the valves or muscle of the heart. A second (Case 4) had a spontaneous left pneumothorax and here again small QRS waves and abnormal T-waves were present. The T-waves in Leads I and II rise

immediately from the S-wave without an intervening iso-electric level (Fig. 14). In fact these changes were reported as evidences of myocardial disease although on x-ray examination the heart appeared normal, although the aortic arch shadow was widened (Fig. 13).

One patient (Case 5) presented a spontaneous right pneumothorax. The electrocardiogram showed a simple tachycardia, small T-waves,

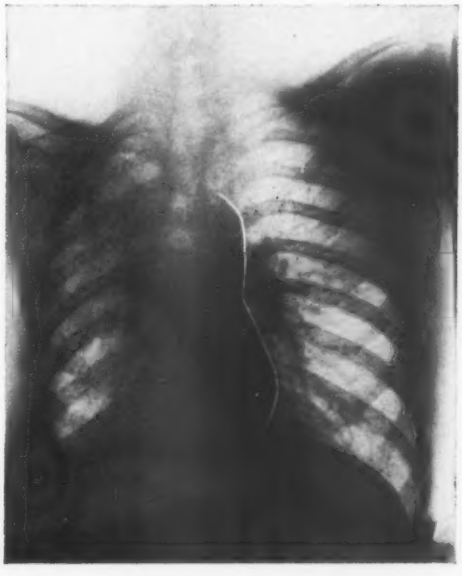

Fig. 10.—Case 2. Roentgenogram taken six months later, no pneumothorax. Fibrotic changes in right lung pulling mediastinum to right.

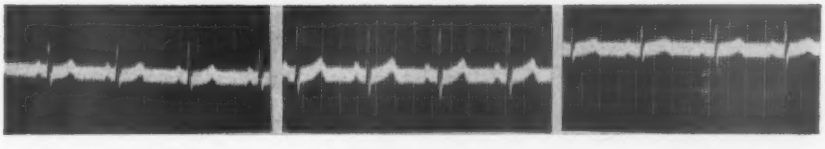

Fig. 11.—Case 2. Corresponding electrocardiogram; i.e., normal one.

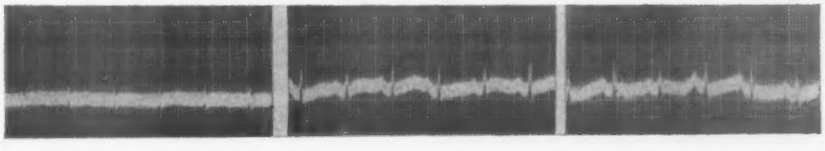

Fig. 12.—Case 3. Electrocardiogram from case of spontaneous right pneumothorax. A tachycardia, low voltage of the QRS group and T-waves, slight right axis deviation of the QRS group.

right axis deviation, and an occasional ventricular premature contraction. The T-waves were reported as suggesting poor contractile force of the heart muscle (Fig. 15). The x-ray record showed no abnormality of the heart except perhaps that it was slightly displaced to the left.

Case 6, a patient with a long-standing spontaneous, idiopathic left

pneumothorax, gave an electrocardiogram with a moderately small QRS group and abnormal S-T segments suggesting myocardial involvement (Fig. 17). The x-ray film revealed only a calcified aortic knob (Fig. 16).

Case 7, a patient with an artificial right pneumothorax, in which only 500 c.c. of gas were injected (Fig. 18), showed a slight right axis devia-

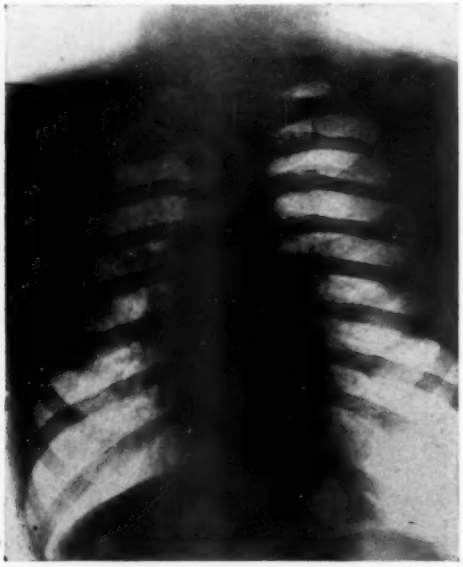

Fig. 13.—Case 4. Roentgenogram showing spontaneous left pneumothorax.

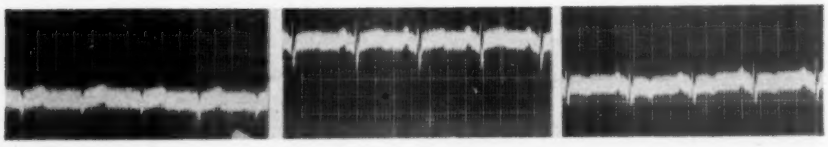

Fig. 14.—Case 4. Electrocardiogram showing small QRS waves, right axis deviation of the QRS group and abnormal S-T segments.

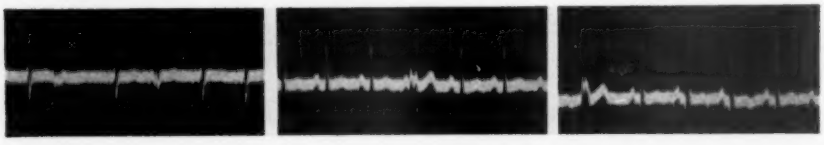

Fig. 15.—Case 5. Electrocardiogram from case of spontaneous right pneumothorax. Simple tachycardia, small T-waves, right axis deviation of the QRS group, and an occasional ventricular premature contraction.

tion (Fig. 19). This patient was reexamined on October 21, 1927. The x-ray film showed normal lungs and no air in either cavity (Fig. 20), and the electrocardiogram was now normal, i.e., it showed no evidence at all of a right axis deviation, no S-wave present in Lead I, QRS group in Lead III much smaller, and the T-wave in this lead now inverted; in other words, an entirely different electrocardiogram (Fig.

21). *The two x-ray films (Figs. 18 and 20) seem to prove definitely that the pneumothorax had produced rotation of the heart, for in no other way can one account for the difference in the shape of this organ.*

DISCUSSION

Before an analysis is made of the previously mentioned observations, it must be remarked that the electrocardiograms were always taken in the same position, i.e., in the recumbent. The influence of digitalis was excluded.

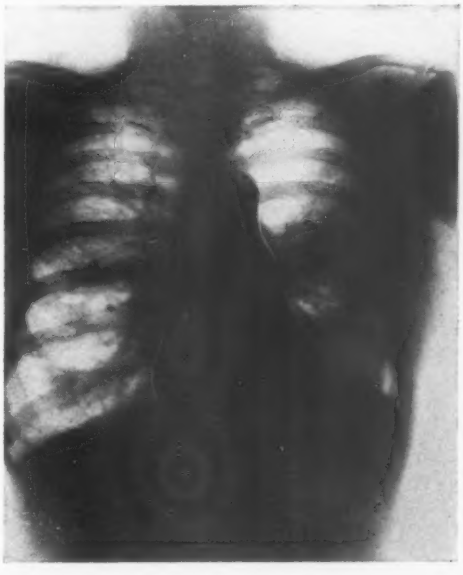

Fig. 16.—Case 6. Roentgenogram from long standing case of idiopathic left pneumothorax.

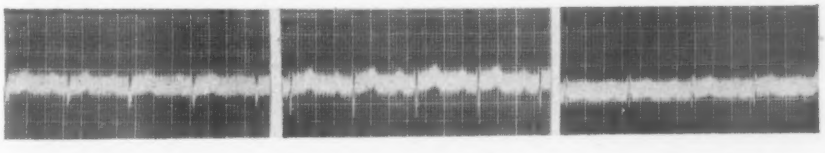

Fig. 17.—Case 6. Electrocardiogram showing moderately small QRS group and abnormal S-T segments.

The commonest electrocardiographic finding in the seven patients was the deep S-wave in the first lead. All seven cases revealed this change. A right axis deviation of the QRS group (right ventricular preponderance) was next most common, being present in five cases, i.e., in all the pneumothorax cases involving *the right side.* One would expect these two findings to appear together because an S-wave in Lead I is a sine qua non for a right axis deviation. A low voltage of the QRS group was present four times; small T-waves three times—always found with a small QRS group; abnormalities in the shape of the T-waves and S-T intervals—changes commonly associated with

myocardial involvement—were present three times. In three other cases these latter deviations were suggestive of myocardial involvement. Although a tachycardia was present three times, it is probably not characteristic.

Clinically these changes are important because they are similar to some of those usually tabulated for myocardial disease, e.g., the small QRS and T-waves and the S-T changes. Two of the patients who showed these changes proved to have normal hearts at autopsy. In pneumothorax, therefore, the diagnosis of myocardial disease must be

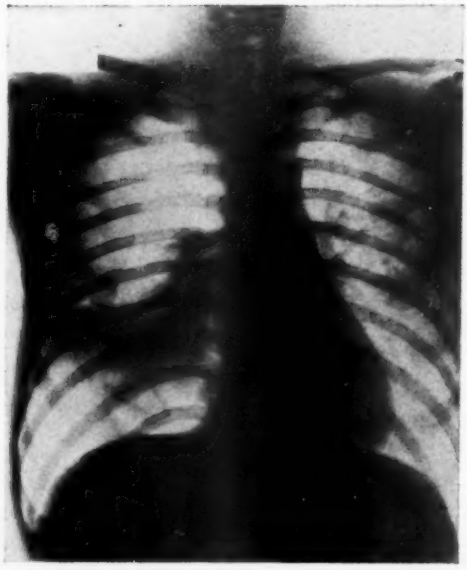

Fig. 18.—Case 7. Artificial right pneumothorax. Roentgenogram taken after an injection of 500 c.c. of gas.

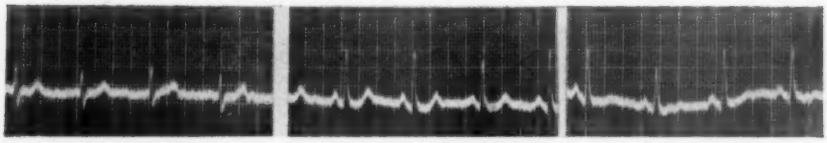

Fig. 19.—Case 7. Electrocardiogram showing slight right axis deviation of the QRS group.

made warily, if at all. The right axis deviation is very significant. It seems to explain the nature of the displacement of the heart in right pneumothorax. A rotation of the anterior surface to the left on a longitudinal axis of the heart or of the apex to the right on an anteroposterior axis would produce such right axis deviation and it is probable that such rotation of the heart accounts for the electrocardiographic changes observed.

Many authors have emphasized the importance of rotation in influencing the form of the electrocardiogram, and Meek and Wilson have

obtained results similar to the foregoing in their experimental rotation of the dog's heart either to the left around a longitudinal axis or to the right on an anteroposterior axis.

In the present series it was the spontaneous pneumothoraces, rather than the artificial ones, that produced the more marked and characteristic changes. Such changes, in the artificial pneumothorax cases, may be most marked four to five days after the pneumothorax has been produced, as is seen in Case 2.

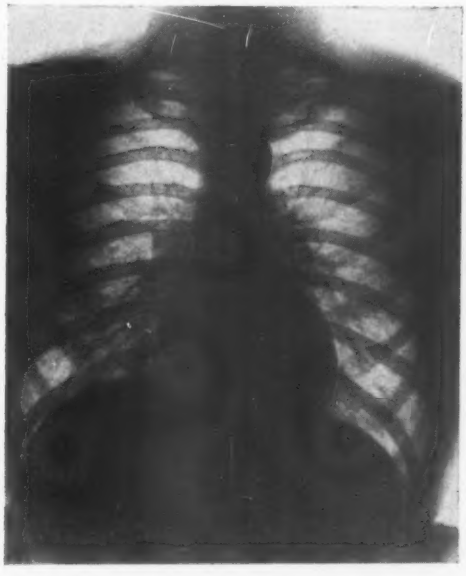

Fig. 20.—Case 7. Roentgenogram showing normal lungs.

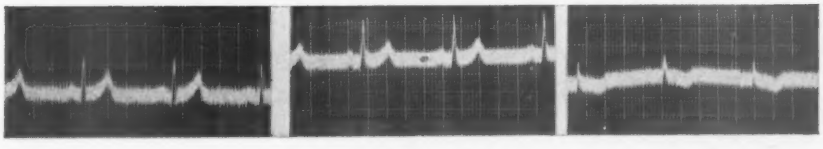

Fig. 21.—Case 7. Normal electrocardiogram, no S-wave in Lead I, QRS group in Lead III much smaller, and T-wave in this lead now inverted.

The S-T changes, usually interpreted as indicating myocardial involvement, require further discussion. These findings have been considered almost specific for coronary artery closure and are particularly to be found soon after an infarct of the heart muscle. The fact that the S-T voltage abnormalities are found also in the rotated heart of pneumothorax cases suggests that a rotation of the heart may occur after an infarct in the left ventricle. It is not illogical to assume that the contraction of the healthy muscle on the one hand, in antagonism to the dead muscle on the other, will produce a torque effect—a rotation of the heart. Hence it is possible that in a myocardial infarct

and in a pneumothorax the same rotation, with the same electrocardiographic changes, may occur.

The low voltage of the QRS group and the T-waves is probably due to change in position of the heart but may be due to the fact that air in the chest conducts poorly the electrical currents in the heart to the surface of the body, i.e., to the leads.

SUMMARY

The electrocardiograms of seven cases of pneumothorax, five spontaneous and two artificial, are described. In three, and possibly in six of the cases, the changes were significant enough to suggest the presence of myocardial involvement, and yet three of these definitely had good heart muscle. A prominent S-wave in Lead I was present seven times—in 100 per cent of the cases; a right axis deviation of the QRS group occurred in five cases, small amplitude of the QRS group four times, and small T-waves three times. Unusually high or low voltage of the S-T segments was present three times; finally, tachycardia was noted three times. All of the right pneumothorax cases showed a right axis deviation of the QRS group.

In pneumothorax, particularly right pneumothorax, a rotation of the heart occurs and probably accounts for the findings described. Another way of stating the same fact might be to say that the right ventricle comes closer to the frontal leads than ordinarily.

Follow-up procedures on two patients, whose pneumothorax had disappeared, showed a return to a normal electrocardiogram, with a disappearance of the characteristic changes observed in pneumothorax.

The most marked electrocardiographic changes occurred with displacement of the mediastinum.

It is suggested that the S-T changes noted in acute coronary closures, i.e., the high take-off of the S-T transition period from the QRS group may be due, at least in a measure, to a rotation of the heart which may occur when a portion of the heart muscle dies.

The low voltage of the QRS group and the T-waves is probably to be accounted for either by the rotation of the heart or by the fact that air in the chest is a poor conductor for the heart current.

I wish to express my thanks to Dr. L. A. Conner and to Dr. Wm. R. Williams for permission to use their cases, and for their kind cooperation and assistance.

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ELECTROCARDIOGRAPHIC CHANGES IN A CASE OF SEVERE CARBON MONOXIDE POISONING*

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ASPHYXIA is the cause of the degenerative changes to which carbon monoxide poisoning owes its seriousness. This statement applies to a great extent, also, to poisoning by the inhalation of automobile exhaust gas, for in the combustion of pure gasoline the only toxic constituent of such gas is carbon monoxide, though when benzol is present in the gasoline used, the amount of this substance in the exhaust gas adds somewhat to its toxicity.[1] Benzol, in acute poisoning, as stated by C.-E. A. Winslow, in his *Summary of the National Safety Council Study of Benzol Poisoning,*[2] is a tissue poison, especially neurotoxic, resulting at first in increased respiratory effort, with thus more rapid intake of carbon monoxide, if the latter be present, and finally in asphyxiation by paralysis of the respiratory center. Not only in the anoxemia of pure carbon monoxide poisoning, but when, as in exhaust gas inhalation, the factor of tissue poisoning may be added, the most specialized tissues suffer first; hence, the damage to the nervous system resulting in coma and respiratory failure. Any damage to the heart would, therefore, be expected to affect the A-V node, the S-A node, or the bundle of His, most likely the first of these. This has been shown to be true in asphyxia by Lewis, White, and Meakins[3] in experiments on cats, and by Greene and Gilbert[4] in humans. Furthermore, any disturbance of conduction occurring below the branching of the main bundle is usually permanent. The electrocardiographic changes noted in the following case are, therefore, unusual in that they seem not to conform with the foregoing two statements.

CASE REPORT

The patient, a young man of twenty-three years, was admitted to the hospital unconscious at 10:00 A.M., August 27, 1926, having been found at 7:45 A.M. lying close to the exhaust of a tractor in which "benzol" gasoline was being used as fuel. It was later learned that he had always been entirely healthy with the exception of having had scarlet fever at the age of twenty years and gonorrhea a few weeks prior to the present illness.

Upon examination at the time of admission, the patient was found to be completely comatose. The pupils were dilated and fixed; the mucous membranes were bright red. There were hot water bottle burns over the abdomen, the result of first aid treatment before admission. The pulse was regular, rate 140, the systolic

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blood pressure 104 mm., diastolic 70 mm., and, according to one examiner, there was an alternating pulse. The reflexes were all normal. The blood was bright cherry red, and the carbon monoxide saturation was 40 per cent.

The patient was given a mixture containing 95 per cent oxygen with 5 per cent carbon dioxide for thirty minutes by a Henderson-Haggard inhalator.[5] During this procedure the respirations became somewhat more rapid and deeper and the

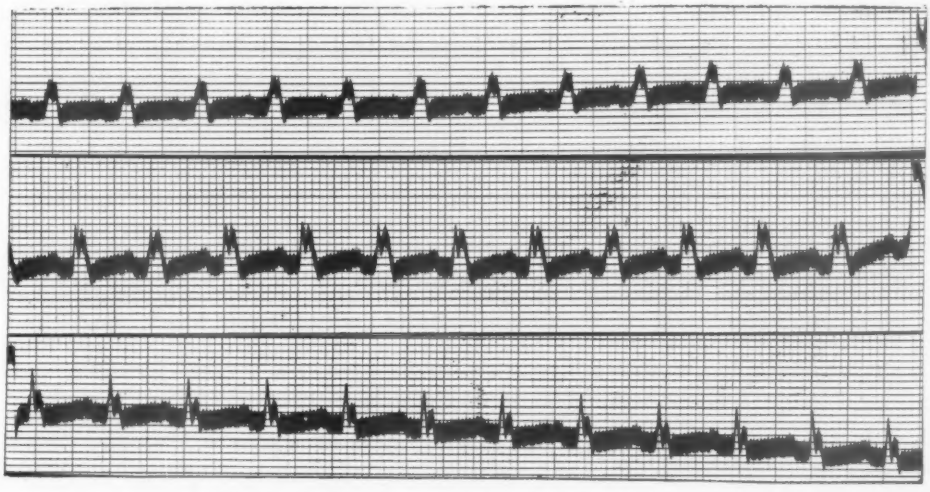

Fig. 1.—Electrocardiogram taken twenty-six hours after the patient was first found in coma.

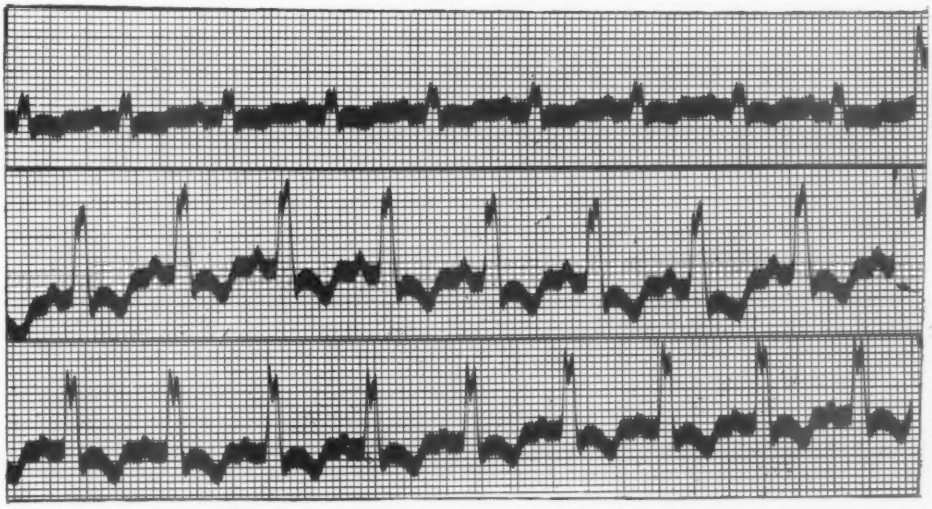

Fig. 2.—Electrocardiogram taken three days after the poisoning occurred.

blood pressure remained constant. The carbon monoxide saturation dropped to 25 per cent. Fluids were forced, and seven hours after admission the patient could be roused. Oxygen and carbon dioxide were again administered during the evening of this first day. There was a slow return to full consciousness, a drop in pulse rate, and intra-ocular movements became normal.

At 10:00 A.M., August 28, twenty-six hours after the patient was found, and twenty-four hours after admission, an electrocardiographic tracing was taken (Fig.

1). This shows sinus tachycardia, low voltage, widened QRS time, notched QRS complexes in all leads, and iso-electric T-waves in all leads. This is the typical curve of conduction disturbance within the ventricles, or so-called intraventricular block. At this time the heart was found to be normal in size, the relative cardiac dullness being 2 cm. to the right in the fourth interspace and 9 cm. to the left in the fifth. There was a tic tac rhythm, and a systolic blow at the apex and in

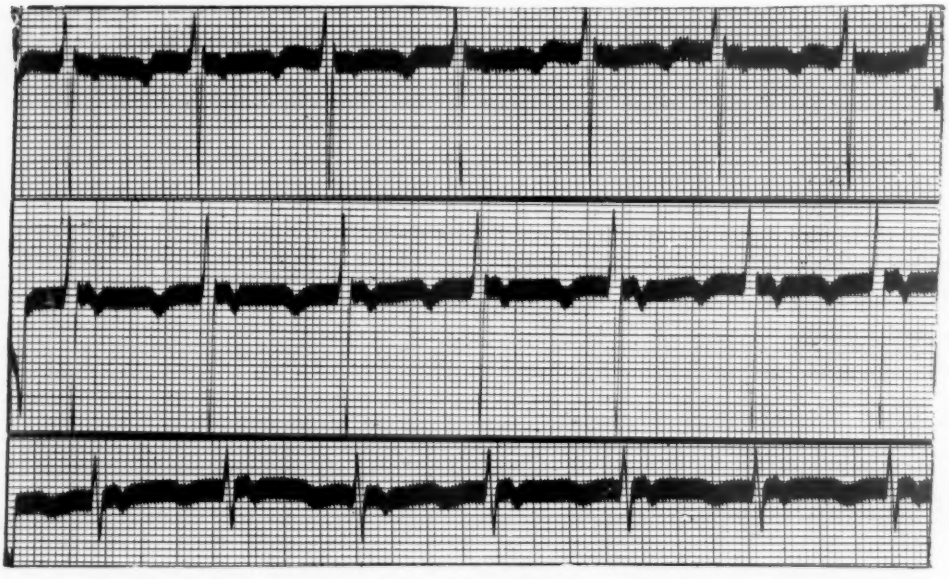

Fig. 3.—Electrocardiogram taken five days after the poisoning occurred.

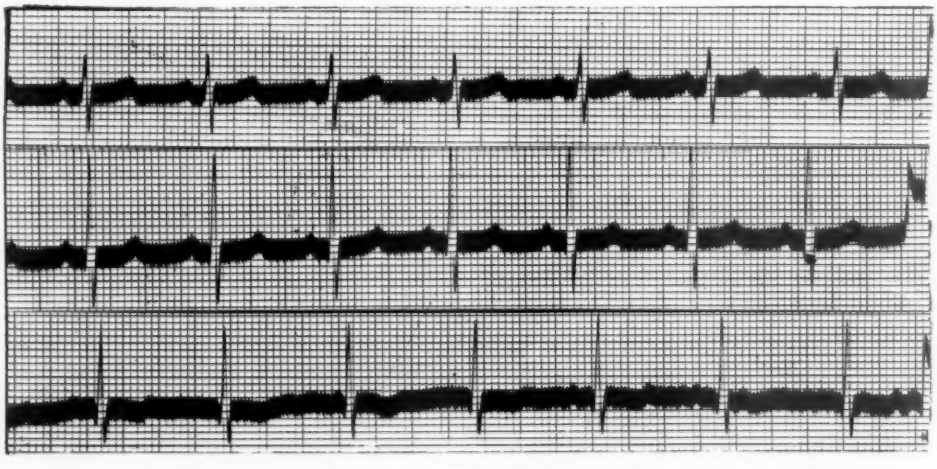

Fig. 4.—Electrocardiogram taken eight days after the poisoning occurred.

the pulmonic area. The radial walls were soft, the systolic blood pressure 88 mm., the diastolic, 60 mm. The lungs were clear, and there was no evidence of congestive heart failure. The carbon monoxide saturation at this time was still 25 per cent. The temperature (rectal) was 102.4 degrees.

On August 30, the patient had improved mentally, though cerebration was still slow. A second electrocardiographic tracing was taken (Fig. 2), which shows sinus tachycardia, widened QRS interval, notched QRS complexes in all leads, and

inverted T-waves in all leads. The voltage has increased considerably over that in the first tracing, but this curve represents, as did the first, impaired conduction in the ventricles. A third electrocardiographic tracing was taken September 1 (Fig. 3), and a fourth, September 4 (Fig. 4), at which time there was found no carbon monoxide in the blood. A final curve was made September 9 (Fig. 5). These three tracings show the QRS time normal, no notching of the QRS complexes, and the T-waves positive in Lead I, and diphasic or inverted in Leads II and III. They are essentially normal electrocardiograms. The blood pressure was 132 mm. systolic and 72 mm. diastolic September 13, and the cardiovascular system was apparently normal in every way. The patient left the hospital November 13, 1926, after rather prolonged surgical treatment of the hot water bottle burns, including skin grafting.

Laboratory Findings.—The urine was not abnormal. The red blood count was 3,840,000; hemoglobin, 83 per cent; white blood count, at the time of admission,

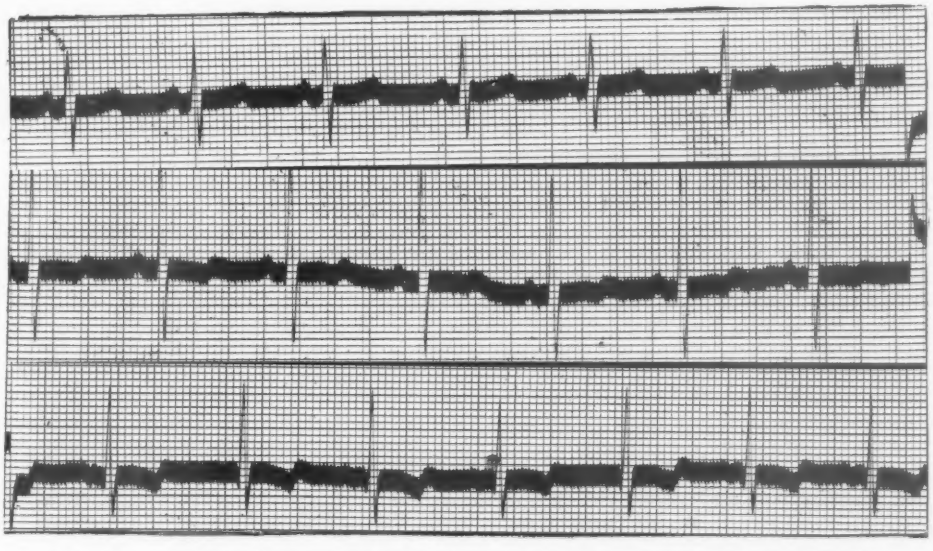

Fig. 5.—Electrocardiogram taken thirteen days after the poisoning occurred.

25,700, with 85 per cent polymorphonuclear leucocytes. By the second day, the white blood count had dropped to 15,300, with 75 per cent polymorphonuclear leucocytes. The blood Wassermann reaction was negative by the Kolmer technic and by cholesterin ice box antigen. The blood chemistry was normal throughout with the exception of carbon monoxide. The phenolsulphonephthalein test, done September 3, showed that 50 per cent of the dye was exereted in two hours.

COMMENT

It would seem, then, that the case above described is that of a young man, always healthy, whose electrocardiogram, taken twenty-six hours after exposure to automobile exhaust gas, suggested temporary damage to the ventricular conducting tissue, and whose cardiovascular findings, clinically and by electrocardiogram, were normal five days after the poisoning occurred.

The literature contains certain reports of electrocardiographic changes in asphyxia due to carbon monoxide inhalation. These have

been experimental. Haggard[6] was able to demonstrate, in dogs, a change in the direction of the T-waves, but nothing else, until the failure of respiration, when increasing degrees of A-V dissociation, even to complete block, occurred. Some of his cases showed ventricular extrasystoles, auricular fibrillation, and finally ventricular fibrillation. He concluded that it required the added anoxemia of respiratory failure to cause these changes. The human experiments, conducted by Greene and Gilbert,[4] and previously mentioned, showed that in the precritical stage there was shortening of the P-R time, and in the postcritical stage a tendency for the A-V node to supplant the S-A node as pacemaker, with consequent slowing of the heart rate, and, later, diminution of conductivity in the A-V node and bundle of His.

Pathological findings, on the contrary, in fatal cases of carbon monoxide poisoning would seem to point to the ventricular muscle, especially the interventricular septum and papillary muscles, as the seat of much damage, as evidenced by hemorrhagic and necrotic foci with surrounding leucocytic infiltration.[7, 8, 9, 10]

SUMMARY

The points of interest in this case seem to be:

1. That the electrocardiographic changes due apparently in the main to asphyxia, but possibly in some degree to the action of benzol, were such as to suggest damage below the division of the His bundle instead of at the A-V node;

2. That these changes, suggesting damage to the ventricular muscle, so rapidly and so completely disappeared;

3. The supposition that if such damage may be done to a presumably healthy heart, an already impaired myocardium might, under similar conditions, suffer irreparably.

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Department of Clinical Reports

DISSECTING ANEURYSM OF THE AORTA COMPLICATING HYPERTENSION*

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IN THE course of a critical analysis of 91 carefully described cases of dissecting aneurysm of the aorta, von Schnurbein,[1] in 1926, found two cases in which hypertension appeared to be the sole causative factor and 17 cases in which it could be regarded as of primary importance. Schilling[2] also had emphasized the part played by increased arterial tension in the production of dissecting aneurysm, and in the older reports, even before the era of the manometer, as in the study of Boström,[3] the hints at vascular strain as an agent in producing aortic rupture are not infrequent.

A characteristic case report follows. It is of a patient whose long-standing hypertension, which had reached an unusually high diastolic level, was terminated by dissecting aneurysm and rupture of the aorta.

Mr. T. H. B., aged sixty-eight years, American, a cashier, came to Dr. W. J. Mallory on Oct. 26, 1927, and was later seen in consultation.

The chief complaint was shortness of breath on exertion, of two months' duration.

Because of this dyspnea, the patient went to Atlantic City about Oct. 15 for a rest, and there, on Oct. 21, had an attack of what he described as heaviness in the chest. There was no pain, but he felt as if he could not breathe, and was unable to speak. This attack lasted over an hour. He spent a day in bed, and was given nitroglycerin by a physician who found the "blood pressure 220." There were no further attacks.

The patient had been free from illness, except for mild digestive disturbances, and an operation, in 1921, for a pelvic abscess, regarded as secondary to appendicitis. At this time, the arterial tension was found elevated.

The work of the patient had been exacting, but without physical strain. His habits were abstemious. He had never had headache, dizziness, or eye symptoms. He had always slept well, and the bowels were kept regular. For a year or more there had been nocturia one time, with occasional swelling of the ankles.

In his family, the patient had no knowledge of circulatory disease. His mother lived to be eighty years old; his father died at sixty-five years.

The physical examination (Oct. 26) showed a well-developed, sparely nourished, elderly man, height 70 inches, weight 133 pounds. The frame was slender, the head long and narrow in type. The color was good.

The pupillary reactions were normal, and the fundi showed only moderate sclerosis of the retinal arteries, without hemorrhage or exudate. The teeth were in repair, the throat was negative, and there were no abnormal pulsations in the neck.

*From the Medical Clinic, George Washington University Hospital.
Presented, in abstract, to the Washington Society of Pathologists, March 3, 1928.

The peripheral arteries were moderately thickened, and the blood pressure was 240/130 mm. Rhythm was regular; rate 80 beats per minute.

The heart gave a strong, diffuse impulse in the fifth and sixth interspaces below and outside the left nipple. Cardiac dullness extended 4 cm. to the right, and 13 cm. to the left of the midline. Percussion did not show increased aortic dullness.

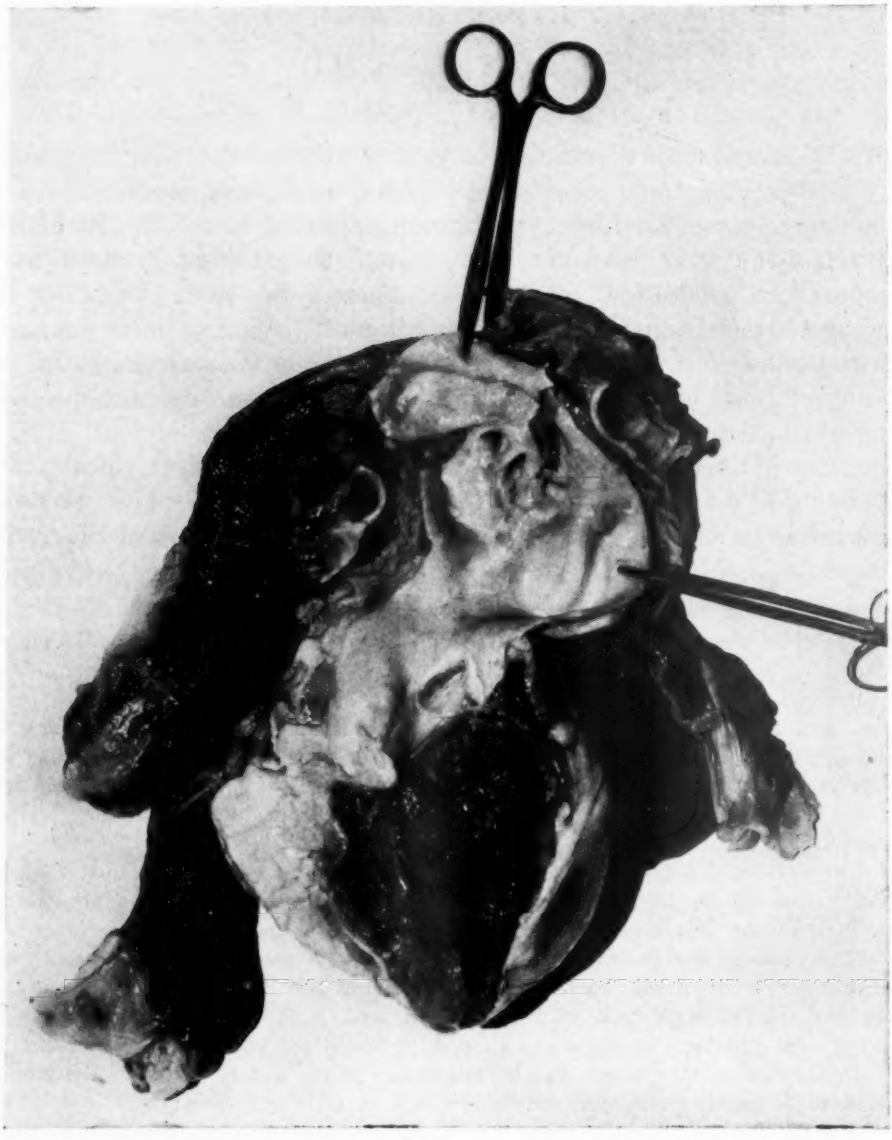

Fig. 1.—Posterior view of heart, with the pericardium reflected. The site of intimal rupture is shown in the arch of the aorta, between, and posterior to, the orifices of the innominate and left carotid arteries. To the left is the blood-filled root of the left lung.

The heart sounds were of fair quality; the aortic second sound was booming. At the aortic area, transmitted down both sides of the sternum, and also heard just outside the apex, was a long, high-pitched diastolic murmur. At the apex there was a systolic murmur.

The abdominal wall was soft, with a midline scar below the umbilicus. There was no tenderness. The liver was not enlarged, and the spleen was not felt.

The knee jerks were present; there was slight edema of the ankles.

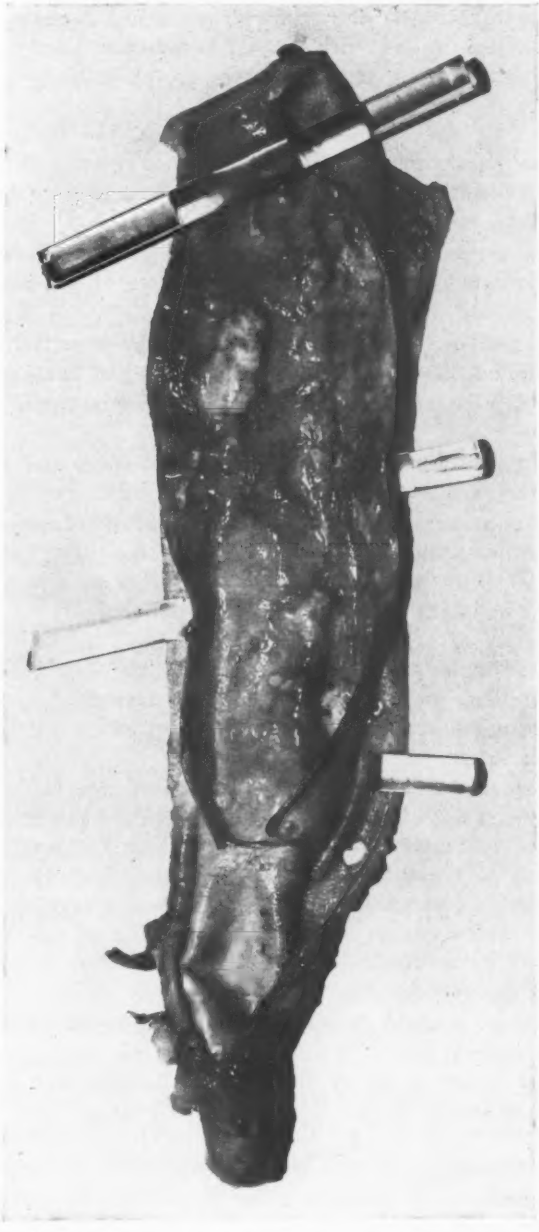

Fig. 2.—The descending thoracic and abdominal portions of the aorta, seen from in front. The "tube within a tube" is well seen just before the bifurcation.

Laboratory examinations showed no albumin in the urine, and no casts. There were a few white, and occasional red, blood cells. A twelve-hour night specimen of urine measured 940 c.c., specific gravity 1.011; the day urine was 455 c.c., specific gravity 1.018. Thirty-five per cent of phenolsulphonephthalein was excreted in two hours. The blood Wassermann and Kahn tests were negative.

Fluoroscopy revealed slight dilatation of the aorta.

The patient was given a belladonna and bicarbonate mixture, remained at work, felt better, and had gained four pounds in weight by Nov. 7, when his blood pressure was 225/140 mm.

On the evening of Nov. 7, two hours after eating a more liberal meal than usual, the patient was seized with severe, continuous pain beneath the upper sternum. It was constricting, radiated across the chest both to the right and to the left, and down the outer (not the inner) side of the arm to the elbow. When the patient was seen three hours after the attack, the thoracic pain was still steady and severe, and there had also developed a constant pain, deep and continuous, in the lumbar region on each side. There was no tenderness in spine or sacroiliac joints.

The blood pressure was 240/130 mm., pulse rate 84, temperature and respiration normal. There was no distention or tenderness of the abdomen.

The patient vomited once, six hours after eating, and half an hour after morphine, gr. ¼, had been given by mouth. Nitroglycerin had been tried by the patient without relief of pain, and during the night another half grain of morphine sulphate, with atropine, was given, with only slight lessening of pain and no sleep.

On Nov. 8, the pain persisted both under the sternum and in the lower back. The arterial tension was 224/130 mm., pulse 80.

On Nov. 9, the temperature rose to 99.5°. The blood pressure was 225/140 mm., pulse 76. Substernal and lumbar pain were definitely less, but in point of degree, and character, the patient now, as on the previous examinations, complained as much of the pain in the back as of that in the chest.

When seen on Nov. 11, the patient had become slightly disoriented. The temperature was 99, pulse 88, blood pressure 190/125 mm., and equal on the two sides. Premature beats were occurring, and the diastolic murmur was distinctly less loud. The pain was still present as a constant ache. In the evening Cheyne-Stokes respiration set in.

On Nov. 12, there was oliguria, and the patient was confused and delirious. There was arrhythmia due to premature beats, and the blood pressure was 200/130 mm. The patient was removed to the hospital at noon. The Cheyne-Stokes breathing continued, and in the evening the blood pressure rose to 230/160 mm., pulse 108. An electrocardiogram showed the presence of numerous nodal and ventricular premature beats. There was no deformity or widening of the QRS group, and the S-T interval and T-wave revealed no evidences of coronary artery occlusion. Digitalis was now given for the first time.

The patient passed a night free from pain, with better orientation. The temperature did not exceed 99.4. The blood pressure on Nov. 13 was 240/165 mm., pulse 108. After three drams of tincture of digitalis, neither the size of the heart nor its auscultatory signs showed obvious change from previous examinations.

At 7 P.M. on this day, the patient was seized with a sharp stabbing pain in the chest, clutched his heart, gasped for breath, and died in a few seconds.

The clinical diagnosis was hypertension, arteriosclerosis, dilatation of the aorta, with relative aortic insufficiency, cardiac hypertrophy, and acute terminal cardiac dilatation.

Necropsy was performed two hours after death by Dr. Tomas Cajigas.

The anatomical diagnosis was dissecting aneurysm of the aorta, beginning with a linear tear in the intima of the transverse portion of the arch, and extending from the first portion of the arch to the right common iliac artery, with rupture externally into the mediastinum and left pleural cavity; hemorrhage beneath

both left and right parietal pleura and the visceral pericardium, and between folds of omentum; dilatation and atherosclerosis of aorta; hypertrophy of the heart; arteriosclerotic kidneys; multiple diverticula of the large intestine.

SUMMARY

After a short period of slight limitation of physical activity, as shown by dyspnea on movement, an elderly man with persistent hypertension, dilatation of the aorta, and cardiac enlargement, and an aortic diastolic murmur, had a brief attack of substernal oppression, followed three weeks later by agonizing substernal pain, which continued for four days without remission and was accompanied by equally severe lumbar pain.

The character and duration of this pain served to rule out the usual forms of angina pectoris; the continued myocardial sufficiency, and the absence of physical, including electrocardiographic, signs discouraged a diagnosis of coronary thrombosis.

At necropsy, the finding of a dissecting aneurysm of the aorta, apparently of several weeks development, judging from the formation of an endothelial lining in the upper part of the new tract, provided an admirable explanation of the clinical course and symptoms. The initial rupture of the intima may be associated with the first attack of substernal distress; the splitting of the media produced the extreme pain of the second attack. Throughout the descending thoracic and abdominal aorta the advancing extravasation of blood brought about, it may be supposed, a rapid dissection with little or transient pain; but the obstruction to the progress of the aneurysm in the region of the bifurcation was productive of the pain in the lumbar region. Finally, the terminal rupture occurred through the adventitia into the left mediastinum, within which the blood was confined until the perforation into the pleural cavity.

From the pathological standpoint, the comment may be added that in the aorta, as elsewhere, sclerotic changes were not marked, and there was no evidence whatever of syphilis. Both of these facts are worthy of emphasis. The later and more marked changes in the development of atherosclerosis, namely, the fatty and calcareous deposits, tend to separate the intima from the media, and an injury to the former is less likely to involve the latter; while in the earlier stage, with what Adami[4] called the "hyaline fibroid" lesions, the connection between the two coats is intact, and an intimal tear is carried through into the media. In regard to the second point, the lamellar arrangement of the elastic fibers of the media is the basis of the formation of the dissecting aneurysm, for the blood finds it easier to separate the weak connective tissue binding these layers together and make its way along the vessel, than to rupture the tough elastic fibers which lie

directly in its course. Syphilitic mesaortitis destroys this orderly arrangement of the medial lamellae, thus preventing dissection, and the syphilitic aneurysm remains localized.

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ANGINA PECTORIS IN A CHILD

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IN a recent paper by White and Mudd[1] attention was called to the comparative rarity of angina pectoris in young people. This was emphasized by their many references to the literature. Because of this dearth of reported cases of angina pectoris in the young, I am recording the following case:

S. S., a female school child, aged ten years, was examined July 1, 1927.

Diagnosis: Postrheumatic heart disease, mitral insufficiency, mitral stenosis, and aortic insufficiency; possible subacute rheumatic fever (because of occasional febrile periods); angina pectoris (the last diagnosis was added August 2, 1927).

Past History: At the age of seven years the patient had rheumatic fever for one month. Since then she has had occasional attacks of tonsillitis and several attacks of joint pains with fever lasting for several days. Recently she has had choreiform movements. She had scarlet fever and measles in infancy.

Present Illness began about three years ago with several nasal hemorrhages. She has had one or two nosebleeds each week since then. She has noted palpitation of her heart, dyspnea, and fatigue on moderate exertion. On several occasions, after particularly severe nose bleeds, she has fainted. At this examination the patient stated that she had never had any pain in her heart. The family history was entirely negative.

Physical Examination: The patient is somewhat undernourished and pale. She is neither dyspneic nor cyanotic. The eyes and nose are grossly negative. The tonsils are enlarged and evidently infected. There is a marked collapsing pulsation in the carotid, brachial, and radial arteries. The precordium shows a well-defined heave. At the apex a systolic and a diastolic thrill are palpable. The heart is enlarged, measuring 3.5 cm. to the right and 9 cm. to the left of the midsternal line and 3.5 cm. under the manubrium. At the apex systolic, presystolic, and diastolic murmurs are heard, and at the base, systolic and diastolic murmurs. The sounds are of fairly good quality. The ventricular and pulse rates are equal (108 per minute), and the rhythm is regular. The abdomen is negative. There are no signs of congestive failure. Blood pressure 112/40. Temperature 99.6°. The electrocardiogram taken July 7, 1927 showed normal sinus rhythm, rate about 100; R_2 quite tall; normal T-waves.

Subsequent Course: On August 2, 1927, the patient was seized with severe precordial pain extending into the left side of the neck and left cheek. The pain was intense and later described by the patient as "cutting." This persisted with short periods of remission for two hours, at which time I saw her. Her face was flushed and damp with perspiration. Occasionally she would scream for an instant and clutch at her chest. Her blood pressure was elevated to 144/40. Her pulse rate ranged from 120 to 130. Because of the severe pain, she was given morphine sulphate, gr. 1/8, and nitroglycerin, gr. 1/200, by hypodermic injection. She was relieved very quickly. The following day, she felt well, and her blood

[1]White, P. D., and Mudd, S. G.: Angina Pectoris in Young People, Am. Heart Jour., 1927, iii, 1.

pressure had dropped to 106/40. On August 9, she had another attack, which closely resembled the first one. The pain was as severe, was situated under the sternum, and radiated into the neck. At the end of one-half hour nitroglycerin, gr. 1/100, was administered under the tongue. The pain was promptly relieved. By October 25, attacks of pain had become rare, were much milder in type, and were all quickly controlled by nitroglycerin. She had had in almost three months a total of three severe attacks and about four much milder ones. They were all similar in location and were all very promptly relieved by nitroglycerin. By November 17, the patient's tonsils had been removed, and she had had no cardiac pain that month.

The case here presented shows, I believe, the characteristic picture of angina pectoris in a child. Physical exertion excited some of the attacks. Mental excitement, such as fear of the consequence of an unprepared school lesson, was also capable of initiating an attack of pain. It is interesting to note that this patient, like those reported by White and Mudd, has aortic insufficiency.

Erratum

In the article, *Pulse Rate Studies with the Pulse Resonator* by Nemet and Boas, which appeared in the last (February) issue of this Journal, the two illustrations on pp. 362 and 363 were misplaced. The illustrations should be transposed so that the one on p. 363 would appear above the legend on p. 362.

Department of Reviews and Abstracts

Selected Abstracts

Rhoades, C. P.: Vegetative Endocarditis Due to The Meningococcus. Am. Jour. Path., 1927, iii, 623.

A case of vegetative and ulcerative endocarditis due to the meningococcus is reported and the literature reviewed.

The identity of the organism is proved culturally and serologically.

The patient was a negro male, 21 years, who was admitted to the hospital with fever, dyspnea, palpitation, cough and chills. He also suffered attacks of precordial pain. Microscopically, the heart showed many endothelial leucocytes between the muscle fibers. There were a few areas which showed necrotic bundles of collagen fibers surrounded by endothelial leucocytes strongly suggestive of Aschoff bodies. In certain areas there was fairly extensive necrosis of muscle fibers with replacement by granulation tissue. Masses of Gram-negative diplococci could be found in the fibrin layers or the valves.

Reid, William D., and Kenway, Florence L.: The Value of the Electrocardiogram in Acute Rheumatic Fever. New England Jour. Med., 1928, cxcviii, p. 177.

For the purpose of determining the value of the use of the electrograph in acute rheumatic fever, the authors have taken a total of 281 electrocardiograms on a series of 26 patients ill with rheumatic fever in the wards of the Boston City Hospital.

Changes indicating involvement of the myocardium were detected in all of the patients. One or more of the three following changes were noted:

(1) Increase in the auriculoventricular conduction time.
(2) Alteration in the ventricular complex.
(3) Changes in the cardiac rhythm.

An increase of the auriculoventricular conduction time was found in 92 per cent of the cases. In 42 per cent it was of the degree found in partial heart-block. Change in the form of the ventricular complex was detected in 80 per cent. The changes included such variations as the presence of an S wave in one curve and its absence in the same individual another day; similar variations though not so marked in regard to the Q-wave; variations in the relative amplitude of the QRS waves; decided variations in the form of the R-wave; variations in the size, extent and direction of the T-wave; and changes in the direction of the S-T portion of the string shadow.

Extrasystoles were found in 34 per cent.

At the time of discharge from the hospital, the changes had disappeared completely in 15 cases. In eight a slight amount persisted and in the remaining three, the new findings were still definitely present. The high incidence of the electrocardiographic signs adds important confirmation to the opinion of many authorities that the heart is involved, though not necessarily to a degree that presents clinical signs, in all cases of acute rheumatic fever.

The finding of evidence of myocardial involvement in electrocardiograms taken in acute rheumatic fever is of clinical importance if the changes are marked or

persistent. Their detection gives reason for continuing to treat the patient, even though he may appear clinically to have recovered.

Feil, Harold and Siegel, Mortimer L.: Electrocardiographic Changes during Attacks of Angina Pectoris. Am. Jour. Med. Sci., 1928, clxxv, p. 255.

Four patients suffering from attacks of angina pectoris presumably due to coronary sclerosis were observed during paroxysms of pain. In three of these patients, there was inversion of the S-T portion of the curve in Leads I and II, while the patients complained of substernal pain. The inversion was greatest in Lead II. The curves resumed their previous contours after the pain subsided. One patient showed no electrocardiographic change during the paroxysm of pain.

Fried, Jacob.: Encapsulated Pericardial Effusion. Am. Jour. Med. Sci., 1928, clxxv, p. 331.

The authors report four cases of rheumatic heart disease in which there was clinical evidence of a fibrinous pericarditis, and in addition, signs of unilateral enlargement of the heart. Further, in corroboration of the clinical signs there was in each case a unique change in the configuration of the heart which they have interpreted as a localized pericardial effusion.

None of the patients died during the course of the study.

The authors summarized the salient points in the diagnosis of this rare condition as follows: (1) The presence of a pericardial murmur. (2) A rapid increase in the cardiac dullness to the right or to the left infraclavicular region. (3) A unique Roentgen configuration of the heart corresponding to the area of dullness. (4) The rapid change in the contour of the heart which would be inconsistent with other cases for such a change in its configuration.

Ghrist, David G., and Brown, George E.: Postural Hypotension with Syncope: Its Successful Treatment with Ephedrin. Am. Jour. Med. Sci., 1928, clxxv, 336.

This report is based on two cases observed in the hospital and carefully studied from the physiological standpoint.

The first patient age 44 years presented the characteristic signs of postural hypotension with syncope: Decrease in the blood pressure to shock level with assumption of the erect position, geographic absence of sweating, failure of the pulse rate to vary with the low blood pressure, with slight depression of the basal metabolic rate, and signs of slight and indefinite changes in the nervous system. A gastric ulcer was present. In this case the use of epinephrin and pituitrin was of no therapeutic value.

The second patient, a man, age 41 years, exhibited the characteristic symptoms and signs described above. It was noted that following the injection of epinephrin the pulse rate increased while with atropine no change in the pulse occurred. There was no increase of blood pressure with epinephrin while the patient maintained the supine position. Following a period of standing the pulse rate further increased to 120 and the systolic blood pressure increased but the diastolic pressure remained as it had been when the patient was lying down. The rise in the systolic pressure was undoubtedly due to the increase in pulse rate. The diastolic blood pressure was not significantly changed by this drug. Epinephrin was most beneficial when given in hourly doses. The pulse rate was increased and maintained.

The authors summarized their study by stating that postural hypotension with syncope probably represents a distinct clinical syndrome characterized by reduction

of the systolic and diastolic pressure to shock level on the assumption of the upright position. The essential disturbance in this disease is the lack of resistance in the splanchnic vessels to shifts in the blood mass and absent or diminished vagus regulation of the heart rate to changes in the blood pressure. The opinion is advanced that this disease represents a hypotonic state of the myoneural structures of the sympathetic and parasympathetic nervous system of unknown origin. The successful treatment by ephedrin in one case allows hope that this drug exercises a specific alleviative effect on the disease.

Wolferth, Charles C.: The Therapeutic Use of Digitalis. Am. Jour. Med. Sci., 1928, clxxiv, 669.

The author describes briefly as part of the symposium on digitalis the therapeutic effects of the drug (1) in the treatment of abnormal rates and rhythms and (2) the treatment of heart failure with normal type of cardiac mechanism.

White, Paul D.: The Clinical Significance of Apical Heart Murmurs. Am. Jour. Med. Sci., 1928, clxxiv, 731.

An analysis of 1050 unselected private patients with apical systolic murmurs seeking medical advice because of cardiac symptoms or signs, has shown that organic heart disease was present in 76 per cent. Of this number in 50 cases, the murmur was transmitted from the base. Sixty-three cases were doubtful and in only 190 was the heart apparently not the site of structural disease. The louder the murmur the greater is the probability of organic heart disease and the worse the prognosis. Valvular disease, cardiac dilatation, or both, appear to be responsible for the bulk of apical systolic murmurs in this group.

The organic mitral systolic murmur is compatible with a long active life but usually is a sign of important heart disease when found clinically. Temporarily its presence need not prevent the demonstration of vigorous cardiac vascular and general strength as indicated for example by various functional tests but eventually it usually represents trouble which limits both duration of life and activity.

In the author's series of 270 cases with very loud mitral systolic murmurs, 75 have died within a very few years of the finding of the murmur. Of the 240 cases with moderate mitral systolic murmurs, 41 have died, while 47 of the 490 cases of slight systolic murmurs have died.

An analysis of 250 consecutive private patients with mitral diastolic murmurs has shown that true mitral stenosis is probably present in about 90 per cent. The balance being due to left ventricular variation, associated with aortic regurgitation or other conditions. Transmission of aortic diastolic murmurs to the apex occurs in about 37 per cent of the cases showing such murmurs.

Organic heart disease is frequently found with no apical systolic or diastolic murmur, as shown by 492 of the present series of 1359 cases of organic heart disease and also quite often, with no heart murmurs at all as shown by 421 of these same 1359 cases. The organic cases without heart murmurs are as important prognostically as those with murmurs.

Simon, Saling and Baum, Felix.: Electrocardiographic Studies in Pulmonary Tuberculosis. Am. Rev. Tubere., 1928, xvii, p. 159.

The authors have studied the electrocardiograms in 250 cases of chronic pulmonary tuberculosis. A right ventricular predominance (right axis deviation) was present in 10 per cent of the 250 cases. A left ventricular predominance was also present in 10 per cent of these cases. These figures do not differ essentially from

those noted by other authors in normal individuals. The low potential sometimes found in Lead I and III especially the former, points to the presence of drop heart.

The authors have studied the amplitude of the T-wave. They find that this deflection is on the average lower than that obtained in normal cases. They assume that the height of the T-wave is determined by the degree of intraventricular pressure and explain the lesser value in these tuberculous patients as due to impairment of cardiac muscle tone.

The authors conclude that the electrocardiogram is of value in pulmonary tuberculosis, giving information in regard to the heart not otherwise obtainable with the same accuracy. The diagnosis of cardiac lesions combined with pulmonary lesions is simplified. Collapse treatment does not always influence the electrocardiographic picture.

Pappenheimer, A. M., and Von Glahn, William C.: Studies in the Pathology of Rheumatic Fever. Am. Jour. Path., 1928, iii, 583.

The authors report their findings in two cases of rheumatic fever presenting unusual cardiovascular lesions. The features of interest in brief are an aortitis of peculiar characteristics not heretofore described and a specific type of panarteritis affecting the larger arteries.

In the aorta there were found abundant lesions in the outer portion of the media following the distribution of the vaso vasorum. In addition there is a pathological process affecting predominantly the intima and subjacent portions of the media. This process can be easily differentiated from the common form of arteriosclerosis.

Case 1 was that of a white male, 15 years old, who died after rheumatic carditis. The heart showed the usual characteristic findings in the valves, myocardium, endocardium, both in the ventricles and in the auricle.

The authors' description of the gross and microscopic changes is as follows: In each of the sinuses of Valsalva are small ridges or plaques in the intima. These are pale brownish in color, glistening and translucent. Above the junction of the anterior and left posterior leaflets of the aortic valve is a larger similar plaque which reaches to the orifice of the left coronary artery. Above the anterior and right posterior aortic leaflets, and separated from them by apparently normal aorta, is a brownish, translucent intimal plaque having a delicately ridged surface as though small areas had become confluent. In nearby portions of the intima are several small, rounded, or oval areas of similar appearance. Near the margin of the large plaque are a number of narrow atheromatous streaks; these are opaque, orange-yellow and dull. The pale brown elevations are very similar in appearance to the lesion of acute rheumatic auricular endocarditis, and are in sharp contrast to the dull opaque orange-yellow atheromatous lesion, and the grey glistening wrinkled plaque of syphilitic aortitis. Because of the restrictions imposed upon the necropsy, the remainder of the thoracic aorta was not removed. In the abdominal portion are a few characteristic atheromatous streaks. Near the bifurcation the wall of the aorta seems distinctly thickened as compared with the vessel wall in the region of the diaphragm.

A block was taken from a translucent plaque from the ascending aorta. Here are found bands of nonnucleated fibrillar material in the intima, bordered by rows of deeply staining cells with basophilic cytoplasm and one or occasionally two large vesicular nuclei containing a central dense clump of chromatin. In their arrangement and staining reaction, these cells are identical with those so characteristically present in the auricle. Where these cells are more sparsely arranged, they assume very irregular shapes. The nucleus is often lobate or bent upon itself, and long plasmatic processes can be traced from the cell body into the crevices of the fibrillar matrix.

The medial lesions in the areas below the intimal plaques are diffuse, but definite. They are best brought out in sections prepared by the Weigert-Van Gieson methods. Perhaps the most striking change is complete loss of muscle cells over large areas, so that only the collagen and elastic fibers persist. This change is foreshadowed in the hematoxylin-eosin preparations in which the muscle nuclei in the corresponding areas have already disappeared. The elastic fibers are thinner, less wavy and less well stained than in the better preserved regions of the media. In places there is actual rupture or fragmentation.

Toward the outer third of the media are encountered perivascular lesions which are like those described in previous papers. They are unusually numerous and many of the cells found in the vicinity of the nutrient vessels have as definitely the character of "Aschoff cells" as in any case which we have studied.

Blocks taken through the large plaques above the aortic leaflets bring out certain additional features. The intimal plaque itself is composed of a rather loose tissue made up of irregular cells tending to a vertical orientation and separated by an indefinite fibrillar stroma. At the margin of the plaque, the endothelium, which in the normal portion of the aortic wall consists of a flattened layer of single cells, becomes dissociated; the individual cells assume a polyhedral form and appear to migrate into the subjacent stroma. Occasionally clefts containing red blood cells are bordered by such polyhedral cells. In certain sections, the clefts appear to communicate with the lumen of the vessel.

This peculiar tissue resembles in no wise the tissue which composes the ordinary atheromatous plaque. Sections stained with Scharlach R show only occasional cells containing finely divided fat which with the polarizing microscope is not anisotropic. There are no free lipoid masses, no large foamy fat-containing cells and no cholesterin crystals. In one field the internal elastic lamella is split into two distinct layers; the upper one is intact, lying immediately beneath the stratum of intimal tissue described. The lower lamina in places is interrupted by patches of scar tissue, about which are grouped irregular vertically disposed cells. This gives indisputable proof of the cicatricial character of these intimal lesions.

The adventitia is very greatly thickened in all blocks examined, and particularly in those taken from the ascending arch in the region of the plaques, and from the lower abdominal aorta. The nutrient arteries have thick walls; the different coats are not easily distinguished. The lumen is often narrowed and in some of the large vessels lined with a layer of new tissue closely resembling that in the intima of the aorta. The elastic fibers are frayed and reduplicated and the muscle fibers are separated by connective tissue and young fibroblasts.

In one nutrient vessel in the abdominal portion, there is a focal collection of large cells resembling an Aschoff nodule.

Similar changes to a lesser degree were observed in the coronary vessels, the arteries of the celiac plexis, and the superior mesentery and renal vessels.

The lesions which have been depicted in the aorta are, in these cases, not mere extensions from the arotic valves, since they occurred at a distance. The aortitis is of two types which may be associated or occur discretely and which probably corresponds to the route of infection. When this takes place from the lumen, there is produced a diffuse reaction in the intima and subjacent media, comparable to that which so frequently affects the auricular endocardium. On the other hand, infection reaching the aortic wall via the vaso vasorum produces a focal perivascular reaction, more closely analogous to the Aschoff nodules about the small branches of the coronary vessels. It is not maintained that this difference in type of reaction is of fundamental importance; it may be in part dependent upon the pecularities of the structural arrangement of the tissue in which the reaction is occurring.

It is of importance, however, to recognize the fact that the pathology of rheumatic infection does not begin and end with the Aschoff nodule. Experience shows that it is necessary to recognize as equally distinctive the more diffuse reaction where it occurs in the valves, endocardium, aorta or smaller visceral vessels.

Morawitz, P. and Hochrein, M.: On the Diagnosis and Treatment of Coronary Sclerosis. München. med. Wchnschr., 1928, v, 17.

During one year, of 137 autopsies, where essential changes of the myocardial and the coronary vessels were present, 91 showed a coronary sclerosis, and 46 presented merely myocardial changes without coronary sclerosis. Of these 91 cases, 75 per cent did not show subjective symptoms of heart disease. Changes in the aorta were present in only two cases, so that the authors do not think that angina pectoris is due to aortic disease.

The authors discuss the discrepancy between the classical and anatomical findings. If the classical symptom complex of angina pectoris was present, sclerosis of the coronary vessels at their origin from the aorta was found at autopsy. Only very minor changes of the aorta could be found. The authors think that they can assume, with great probability, the presence of coronary sclerosis clinically, if in persons of advanced age, signs of heart disturbance develop, even if such are unaccompanied by pain. The diagnosis of coronary sclerosis in such cases seems of importance for the prognosis, because every patient of this kind is threatened by sudden death.

The objective signs of the prognosis are the changes of the blood pressure during the attack, the effect of vagus pressure, pulse irregularities and a high venous pressure. For the therapy, besides general dietetic measures, the authors recommend the use of digitalis and salyrgan.

Moritz, F.: On the Diastolic Capacity of the Heart Chambers. München. med. Wchnschr., 1928, v, 21.

Experiments were done to ascertain the rôle played by increased diastolic volume of the heart in changing the minute output. The method of Hesse was used. Dogs' hearts, kept alive in Ringer's solution at body temperature, were subjected to different pressures of Ringer's solution, and the changes in heart volume noted. Curves are included showing the changes in volume accompanying the different pressures applied. It is impossible to determine directly these changes in human hearts by the same method, as an indirect method was used. The capacity of hearts was determined by means of wax models of the cardiac chambers imitating the diastolic filling and systolic contraction. In this way considerable increase of the capacity was possible.

Edmunds, C. W., and Johnston, B. S.: The Circulatory Collapse in Diphtheria, Jour. Am. Med. Assn., 1928, xc, 441.

The most frequent cause of death in diphtheria is circulatory collapse. This collapse may be the result of the effect of toxin on the heart muscle and its mechanism or there may be collapse due to failure of vasomotor response in the systemic circulation. There still is no known mechanism by which this failure of the circulation results. It has been pointed out that in these cases the heart and vasomotor center have even shown not to be primarily the cause of circulatory failure.

The authors have studied the condition of the peripheral vasomotor mechanism in an attempt to throw light on this phenomenon. They have pointed out in a previous study that epinephrin partially or completely fails to bring about an

increase in the systolic pressure in these cases of circulatory failure. They have administered epinephrin to dogs poisoned by diphtheria toxin and find no response to the drug. If the test is made early when the blood pressure fall has just begun and the pressure is still fairly good, then the response approaches that seen in normal animals. If, however, the collapse has progressed further, the response is diminished until results of injections of epinephrin are nil.

An important difference in response from that shown in epinephrin is that produced by extracts of the pituitary gland. If they have been preceded by injections of dextrose to give the needed volume of circulating fluid the pituitary extracts usually elicit a very favorable response. Experiments have been carried out on dogs and also on guinea pigs and with the result that the animals respond to high dilutions of pituitary extract.

As a result of these experiments, the authors believe that the receptive substance at the myoneural junction is the part of the nervous mechanism which is attacked by the diphtheria toxin. They point out that it is probable this action is rarely one of complete paralysis. The splanchnic vessels are those most apt to be affected.

For the present, evidence is that the fatal circulation collapse is due primarily to an action of the toxin on the receptive substance of the splanchnic nerve producing a lost control of the abdominal vessel by these nerves. The vessels relax and blood pressure falls resulting in a deficiency in the amount of blood returned to the heart. The general arterial pressure fails also causing an anemia of the medullary centers and producing death.

Levy, Robert L., and Golden, Ross: Some Effects of Roentgen Irradiation in Rheumatic Carditis. Am. Jour. Roentgen. and Rad. Therapy, 1927, xviii, 103.

With the idea of attempting to influence rheumatic lesions in the myocardium, 20 patients with rheumatic carditis were given 139 irradiations to the heart. The machine setting was calculated to yield 1/10 of an erythema dose distributed throughout the heart muscle. Frequent electrocardiograms and numerous teleroentgenograms were made.

Changes in the form of the electrocardiogram were observed in 14 of the 20 cases. The changes were similar to those sometimes found in the course of rheumatic fever and regarded as due to myocardial involvement. It is suggested that the alterations noted following roentgen irradiation of the heart occurred as a result of the action of the rays on the tissues of the heart with modification of the rheumatic lesions.

Of the 20 cases, 17 showed definite improvement. The value of roentgen irradiation of the heart as a therapeutic measure in rheumatic carditis is not stressed on the basis of these observations, because of the small number of cases studied and the relatively short period of follow-up.

In 8 patients, mildly unpleasant symptoms were noted. In two cases of Streptococcus viridans endocarditis, roentgen irradiation of the heart caused no change in the form of the electrocardiogram nor did it modify the fatal progress of the disease.

Nichols, B. H.: Some Observations From a Roentgenological Study of the Aorta. Radiology, 1927, ix, 136.

The first and most important fact pointed out is that the position of the aorta in relation to the heart and mediastinum may vary in normal individuals. This variation in position may be from an extreme right to extreme left position of the ascending portion of the aorta. Depending on this position will be the character of shadow seen on fluoroscopic examination or in films.

If the aorta lies to the right, there may be considerable pulsation of the right mediastinal shadow. This pulsation is probably transmitted from the aorta to the superior vena cava. In these, with aortitis or thickening of the arterial wall, the aortic shadow may be increased in size.

The authors feel the roentgen ray is the most reliable method of early diagnosis of aortitis or aneurysm.

Films made in the posterior anterior position and in the oblique and lateral positions are the most valuable in the diagnosis of early stages of the disease.

Kininmonth, J. G.: The Circulation Rate in Some Pathological States, with Observations on the Effect of Digitalis. Quart. Jour. Med., 1928, xxi, 277.

The author has studied the cardiac output and circulation time in 24 patients, using the ethyl-iodide method of Henderson and Haggard and the apparatus described by Davies and Gilchrist. The author feels that this method is satisfactory and approximates the results obtained by those methods based on the Fick principle.

Six cases of aortic valvular lesions and three cases of mitral stenosis were examined. From the results it would appear that these lesions do not affect appreciably the rate of blood flow provided the myocardium is functioning efficiently.

In three cases of auricular fibrillation, there was no appreciable effect on the cardiac output. However, in the presence of cardiac failure, the circulation rate is low.

Other cases studied show that the state of the myocardium and cardiac efficiency remains whether or not there will be a change in the circulation rate.

Allan, George A.: The Early Detection and Supervision of Rheumatic Infection in Children. Brit. Med. Jour., 1928, i, 39.

In this address the author reviews clearly and at length the attitude that is being adopted toward rheumatic infection in the light of more recent developments in the study of this disease. He points out the importance of caring for those conditions and diseases which lead up to the occurrence of carditis in children.

He points out rightly that unless something be done to prevent the occurrence of these conditions the incidence of the disease will probably continue as high as ever. If attention is directed toward recognizing the early manifestations of the disease little can be accomplished more than to prolong the life of patients suffering with rheumatic carditis. If the heart is involved, the situation is undoubtedly bad.

In some detail he outlines the supervision that should be given these children by practitioners in the home, in the school and the hospital. He emphasizes the long duration of the convalescence and the need for such detailed care.

This address should receive careful attention from all who might be interested in this rapidly increasing disease.